IMPROVING CLINICIAN CARE PRACTICES OF PATIENTS WITH SICKLE CELL ANEMIA: AN INTEGRATIVE REVIEW

An Integrative Review

Submitted to the

Faculty of Liberty University

In partial fulfillment of

The requirements for the degree

Of Doctor of Nursing Practice

By

Tracey Ann Royer

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Lynchburg, VA

July, 2021

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Scholarly Project Chair Approval:

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Vickie Moore, RN, DNP, FNP-C Date

ABSTRACT

Sickle cell disease (SCD) is an inherited hematological disorder that can have a debilitating impact on those affected. The disease is caused by a mutation in the gene encoding for hemoglobin that alters the formation of the red blood cell to a sickle shape, the source of the term *hemoglobin S* or *sickle hemoglobin*. The most common manifestation of this disease state is acute pain due to the effects of vaso-occlusive crisis. The burden of disease can be intense, requiring frequent and prolonged hospitalizations with disruptions in one's quality of life. Despite the gravity of this disease state, there exist pitfalls in clinical practice that can negatively impact the quality of care one receives. Patients with SCD have been subject to negative perceptions and deep-seated biases from varying health care providers or personnel. Contributing to this care experience is providers' lack of thorough understanding of the disease state, training, exposure, and suboptimal racial or cultural competence. As a means of improving care practices of clinicians and their preparedness to care for those with SCD, clinician-targeted sickle cell education has been proposed. Related literature on the subject matter has been explored and offers support for the benefits of the implementation of continued clinician sickle cell education in practice.

Keywords: sickle cell disease, education, care barriers, stigmatization, quality care

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Last but not least, I would like to thank my supportive husband, Ardent, as well as my daughters, Milan and Eryn, for their unconditional love, patience, and support. Additional gratitude goes out to my mother for her continued encouragement over the years.

My experience at Liberty University has been a blessing that I will forever treasure. Through lessons learned in conducting this project, my goal is to advocate for improved patient care practices, promote health care equity, and enhance the overall professionalism of nurse practice.

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List of Abbreviations

Agency for Healthcare Research and Quality (AHRQ)

American Association of Colleges of Nursing (AACN)

A Measurement Tool to Assess Systemic Reviews (AMSTAR)

Centers for Disease Control and Prevention (CDC)

Centers for Medicare and Medicaid Services (CMS)

Cumulative Index to Nursing and Allied Health Literature (CINAHL)

Doctor of Nursing Practice (DNP)

Medical Literature On-Line (MEDLINE)

National Heart, Lung, and Blood Institute (NHLBI)

Sickle cell disease (SCD)

SECTION ONE: FORMULATING THE REVIEW QUESTION

Introduction

Sickle cell disease (SCD) is the most prevalent hemoglobinopathy worldwide. This disease state requires daily care and can potentiate long-term disability. Countries of sub-Saharan Africa have the most occurrences of disease, with Nigeria having the highest prevalence followed by Cameroon, the Republic of Congo, and Ghana (Centers for Disease Control and Prevention [CDC], 2020). Other individuals commonly affected are those of Caribbean, Central American, Saudi Arabian, Indian, and Mediterranean descent, as well as those of Greek, Italian, and Turkish heritage. The World Health Organization has recognized sickle cell disease as being of global health significance and suggested that practices to mitigate poor health outcomes should be implemented for the impacted regions (World Health Organization, 2019).

In the United States, it is estimated that 100,000 Americans will be affected by SCD yearly. Those predominantly impacted are African American, accounting for 1 out of 365 births (Matthie & Jenerette, 2015). The disease affects the ability of hemoglobin to carry oxygen to vital organs due in part to the sickling presentation of the red blood cells (Matthie & Jenerette, 2015). Hemoglobin S polymerization is the pathologic process at the root of the red blood cell sickling. If patients are not treated expeditiously, there will be worsening hemolysis with subsequent vaso-occlusion. Presently, there are no standard treatments to cure SCD. Current therapies are more congruent with supportive care. Though finding a cure has been a slow process, researchers have been investing in new therapeutic agents and have explored bone marrow transplantation as a treatment option (Matthie & Jenerette, 2015). Not all patients will qualify for stem cell transplants, as there are significant risks involved. Donors must closely match for the transplant to be successful. The financial burden of SCD is high for the affected

individual and the health care system. Based on the national data, the estimated incremental economic burden of SCD in the United States is $2.98 billion per year (Huo et al., 2018).

One of the most burdensome manifestations of SCD is pain, which can lead to debilitating effects on the patient's quality of life. In addition, sickle cell anemia has been found to have a negative impact on one's mental health, which can lead to depression and anxiety (Matthie & Jenerette, 2015). Despite what is known regarding SCD, stigmatization exists in practice, which impacts quality care outcomes for patients affected by this morbidity (Brennan-Cook et al., 2018). According to Oyedeji and Strouse (2020) the perceived stigmatization can create barriers to the clinician-to-patient relationship. The negative experiences and dissatisfaction can potentiate treatment nonadherence and, ultimately, adverse outcomes. Aims to improve the clinician care approaches and reduce bias toward the care of patients with SCD would be favorable in achieving best practice.

Defining Concepts and Variables

Stigmatization

Stigmatization involves the display of negative attitudes or discrimination toward others based upon presenting characteristics such as a medical condition, disability, or mental state (Rao et al., 2019). Other often-stigmatized characteristics include gender, race, culture, religion, and sexuality. Research has shown that stigma is one of the main risk factors of poor health outcomes, particularly for those with mental illness and disease states such as sickle cell anemia. In addition, surfaced stigmatization can lead to delays in treatment, as patients may hesitate to seek care and providers may fail to recognize warranted interventions (Rao et al., 2019).

According to Cronan et al. (2016), stigma has become an increasingly significant challenge in today's society. To reduce stigma across a continuum, various parties need to

recognize the negative consequences that can evolve from stigmatization. Behavioral scientists have studied the negative effects of stigma for individuals dealing with varying medical conditions and have found that stigma dramatically impacts one's desire to pursue treatment for their condition; subsequently, this affects morbidity and mortality rates (Rao et al., 2019). According to Rao et al. (2019), stigmatization is a multilevel phenomenon that requires intervention approaches capable of targeting the intrapersonal, interpersonal, community, organizational, and structural levels. First, at the intrapersonal level, the individual living with the stigmatized condition is the focus. Strategies here may include, but are not limited to, self-help, counseling, and treatment (Rao et al., 2019). Second, the interpersonal level entails interventions that cater to the enhancement of care and support with the stigmatized person's environment. Third, the community level focuses on decreasing stigmatization attitudes and behaviors with community groups through educational strategies, contact, and advocacy (Rao et al., 2019). Fourth, at the organizational level, strategies can include training programs and the formulation of institutional policies. Lastly, the structural level entails interventions that focus on establishing legal, policy, and rights-based structures (Rao et al., 2019).

Sociologist Erving Goffman (1963) described stigma in his stigma theory as a situation in which an individual is disqualified from full social acceptance. Influencing the interpretation of many sociologists, he identified three types of stigma: physical stigma, stigma of character traits, and stigma of group identity (Cronan et al., 2016). Goffman recognized that there could be continual shifts with stigmatization, which can arouse concern (Cronan et al., 2016). Through all the shifts in stigma, one category that has been identified as consistently subject to stigmatization is low socioeconomic standing (Cronan et al., 2016). According to Bulgin et al. (2018), the

medical profession has played a key role in promoting stigma, as it can incite social exclusion. Efforts to curtail stigmatization in medical practice should be undertaken.

Stigmatization related to SCD is evident in clinical practice. Current literature has revealed that the stigmatization of this disease state can have detrimental consequences. Stigmatization relating to health status refers to health-related stigma that involves judgment, devaluation, or social disqualification of people based on particular health conditions (Bulgin et al., 2018). Despite SCD being a genetic disorder, deep-seated racism and health care equity discrepancies have hindered appropriate access to care and funding support (Bulgin et al., 2018). One of the main contributors to health-related stigma in SCD revolves around pain management with opioid care. The chronic or acute pain associated with the vaso-occlusive nature of this disease often requires management with opioids, which can bring concern of dependency or abuse amongst some clinicians.

Clinical Education

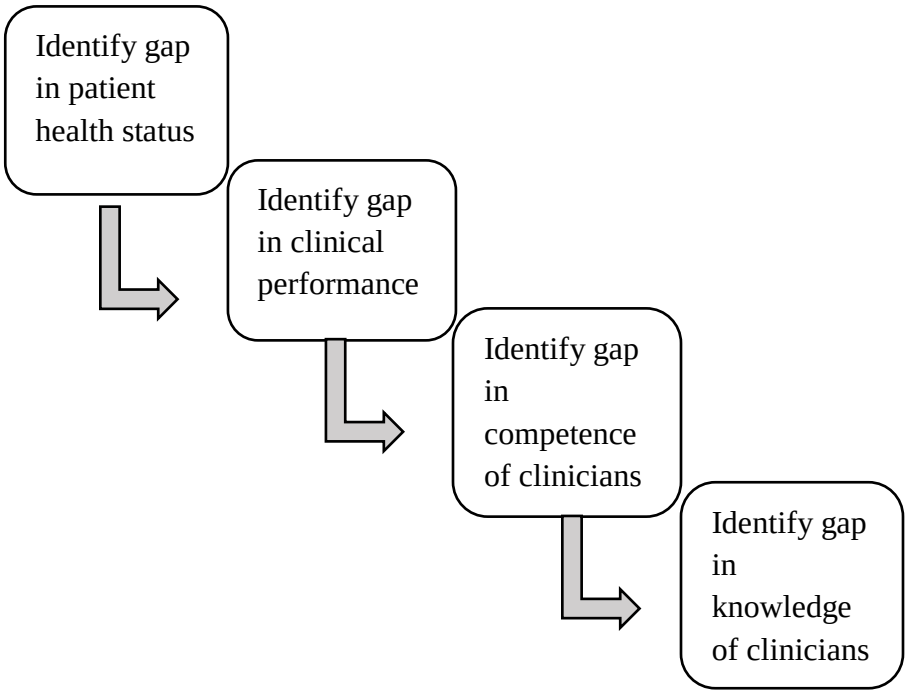
To promote best practice outcomes, the health workforce has a professional responsibility to maintain competency. Through continued education, clinicians can be better prepared to provide evidence-based care for their patients and improve care outcomes. Through education, clinicians can stay current in their field as innovative undertakings, research, and technology enhancements are ongoing. The nurse leader can spearhead initiatives to formulate educational programs for clinicians of their institution. Leadership in nursing is valuable and of benefit to other health care disciplines. The Institute of Medicine noted that nurses should be prepared to lead in all aspects of health care (Joseph & Huber, 2015). Transformational leadership can be used by nurse leaders involved in promoting education for interdisciplinary teams. As an evidence-based theory, transformational leadership is feasible when considering the complexities

of patient care and efforts of collaboration amongst interprofessional team players. This form of leadership can form the basis of practical clinical guidance and education (Joseph & Huber, 2015).

Most health care professionals strive to provide the best care for their patients; however, gaps in care can exist when care provided is not based on the best evidence available today. The use of a conceptual framework to institute educational interventions for clinicians has been advised. The continued professional development framework emphasizes the importance of continuous assessment of areas needing improvement (Moore et al., 2018). Continuous assessment involves three phases: needs assessment, formative assessment, and summative assessment. The needs assessment involves the identification and comparison of what the clinician knows before implementing the learning activity and what they should know or display in order to provide the best care to their patients. In the formative assessment stage, clinicians are monitored as they progress during the learning activity to gauge the decrease in the gap identified during the needs assessment. The summative assessment measures what the individual learner knows and how they perform after participating in a learning activity and compares this information with the knowledge they should have gained to provide quality care to their patients (Moore et al., 2018). To achieve desired outcomes, feedback, an essential part of the conceptual framework, must be given to learners. By receiving feedback, the individual clinician or team can recognize their shortcomings; this will aid in reducing the gaps of care in their practice. Figure 1 highlights the stepwise recognition of gaps that should identify the effective integration of a clinical educational strategy.

Figure 1

Needs Assessment in the Conceptual Framework



Note. Adapted from "A Conceptual Framework for Planning and Teams," by D. E. Moore, K. Chappell, L. Sherman, and M. & Vinayaga-Pavan, 2018, *Medical Teacher, 40*(9), p. 906.

Education and training allow providers the opportunity to improve their knowledge and care approaches for patients with SCD. Decreasing knowledge gaps for the health care providers can likely contribute to improved quality of care and enhance the care experience. Considering the stigma that has been associated with SCD in clinical practice, it is essential for health care professionals to include the patient and their families during the educational initiative. To narrow the SCD knowledge gap, decrease negative perceptions, and gain best practice results, it is crucial for clinicians to gain insight regarding the patients' perspectives of the care they receive.

Quality Care

Quality health is care that is patient centered, effective, timely, efficient, safe, and equitable (Harolds, 2016). These dimensions of quality were explained in the Institute of Medicine's 2001 report, *Crossing the Quality Chasm*. The dimensions need to be addressed to improve health care services delivered to patients (Harolds, 2016). In today's clinical practice, the aims of quality care are continuous. The main focus of health care initiatives are to improve quality outcomes for those seeking care. A shift is currently occurring in practice to value-based care, which has altered the focus of patient care from quantity to quality. In addition to the Institute of Medicine's definition of quality, there have been several established interpretations and strategies of attaining quality from various health care organizations. The Agency for Healthcare Research and Quality (AHRQ, n.d.b.) defined quality health care as doing the right thing for the right patient, at the right time, in the right way to achieve the best possible results. In 2019, the Centers for Medicare and Medicaid Services (CMS) listed six goals of the delivery of quality care, which were published in their *Quality Strategy* as follows:

- Make care safer by reducing harm during care delivery.
- Strengthen the person and family engagement by allowing partnership with care.
- Effective communication should be promoted as well as the coordination of care.
- Prevention and treatment of chronic diseases should be promoted.
- Fostering collaboration with communities to promote best practices for healthy living.
- Efforts should be made to make care affordable.

The nursing profession is obligated to promote quality care for best practice outcomes and to be an exemplar for other health professionals to follow suit. According to the American Nurse Association (2015), nurses should strive to deliver safe, high-quality care to patients in

various clinical settings. The American Nurses Association's (2015) code of ethics states that nurses are expected to carry out their professional responsibilities in a manner that is consistent with the ethical obligations of the profession and quality in nursing care. The nurse leader is prepared through their education and training to spearhead continuous quality improvement initiatives for their organization. Continuous quality improvement is an ongoing process to improve health care outcomes by identifying problems, implementing and monitoring corrective actions, and studying effectiveness (White et al., 2016). Continuous quality improvement involves three significant focus areas: quality assurance, quality improvement, and problem resolution (Johnson & Sollecito, 2020). Quality assurance ensures that an organization meets the requirements for effective health care. Quality improvement occurs when members of the organization work toward better delivery of care for the patients (Johnson & Sollecito, 2020). The goal of problem resolution is for all members of the organization to engage in effective communication strategies for positive collaborative outcomes (White et al., 2016).

It is crucial for clinicians and health organizations to measure their quality care deliverance to determine where they stand along the continuum of care. Through measuring, reporting, and comparing care outcomes, one can also determine whether they are on the path to achieving the Quadruple Aims of Healthcare, which are to improve the patient care experience, improve the health of populations, reduce health care costs, and reduce clinician burnout and dissatisfaction (Jeffs, 2018). Hanefeld et al. (2017) defined outcome measures as changes in the health of an individual, population, or group that is a consequence of an intervention.

National standards and financial incentives have driven the measures of quality. Common analysts of quality measures include the CMS, the Joint Commission, and the National Association for Healthcare Quality. Targets and benchmarks designate how well an

organization's level of quality ranks amongst their competitors. Data on quality measures can be reported as claims, assessment instruments, chart abstractions, or registries (Johnson & Sollecito, 2020). Meeting or exceeding the national targets benefits not only quality of care, but also the organization's financial standing and marketing efforts.

With the aim of quality care delivery, clinicians should integrate evidence-based practice guidelines into their practice all the time. The use of clinical practice guidelines will guide the provider to make appropriate patient care decisions for specific clinical circumstances (White et al., 2016). To assure the gains of quality care, these guidelines should be based on the best available research evidence and practice exposures. Organizations should utilize evidence-based guidelines to guide practice decisions for patients with SCD, especially those with complex circumstances. The American Society of Hematology is a beneficial resource to guide quality care practices for patients with SCD.

Rationale for Conducting the Review

Disease states such as SCD can be challenging to manage for most health care providers, as the burden of disease for the patient can be overwhelming. The primary, debilitating manifestation of SCD is pain. In severe cases when a painful crisis is imminent, these patients may frequently visit the emergency department for pain relief and control of the crisis event. Some providers have referred to patients with SCD as "sicklers," which has been perceived as offensive for many who are impacted by this medical condition (Jenerette et al., 2016). One common perception in practice is that the patient seeking care is motivated solely to receive narcotics. There has been increased associations observed between health care providers and negative attitudes toward those with SCD (Jenerette et al., 2016). Patients with SCD rely on health care providers to believe their report and staging of pain and to recognize signs of

urgency. In the emergency department as well as in the inpatient setting, nurses are often the first clinicians to interact with this patient population. Unfortunately, during their curriculum, nurses receive little education about SCD, impacting the care quality (Jenerette et al., 2016). Likewise, studies have shown that other health care provider disciplines also have had limited focused training with SCD, impacting care outcomes. To improve quality care for those with SCD, it is essential to explore methods to resolve these negative attitudes, as they can influence the provisions of care. Contributing to the negative perceptions is the clinician's lack of understanding regarding the disease burden.

To improve care outcomes, clinician-targeted education regarding SCD and management is proposed. A comprehensive review of literature has been undertaken in an attempt to validate clinician-targeted education as a benefit to practice and overall patient outcomes. The literature was also examined for any specific educational methods and knowledge areas that can be implemented with providers to assist in improving patients' outcomes and decrease the stigma related to SCD.

Purpose

The purpose of this integrative review is to explore and analyze relatable research on how clinician-targeted education can contribute to improving quality care outcomes and decrease stigmatization for patients living with SCD. In addition, by increasing awareness of this practice issue, it is hoped that there will be increased motivation to implement a quality improvement practice change.

Clinical Question

Does clinician-targeted education improve quality care outcomes and reduce the stigmatization of patients with sickle cell anemia?

Inclusion and Exclusion Criteria

The patient population of interest was adults who were between the ages of 18 to 75 years living with SCD globally; however, patients living in the United States were the central focus. All races and gender were included, though African American living with SCD were the most frequent population of focus amongst the studies examined. The pediatric population was excluded from the integrative review in decreasing variability; however, there were some relevant comparisons regarding treatment approaches, particularly during the transition to adulthood. The body of research regarding sickle cell clinician education and its impact on care practices was modest; however, there was extensive literature relating to the effects of clinician perceptions as they pertain to the care of patients impacted by this disease.

The review of literature, based on the guidance of Whittemore and Knafl's (2015) framework, was extensive. Articles and other sources of literary works dating as far back as 1963 were reviewed for the purpose of including relevant historical data. To ensure quality and validity, only peer-reviewed journals were retrieved. Varying study designs were accessed during the review of literature; however, most were descriptive studies. Excluded materials were studies that relied on subjective accounts of clinicians who were not directly involved in the care of patients with SCD. Studies that did not reveal measurable outcomes were also excluded. The majority of the studies used were those incorporating statistical data. Gaining generalizability and avoiding bias was the intent. There will be further discussion relating to search methods utilized.

Conceptual Framework

The integrative review approach is inspired by the conceptual framework of Robin Whittemore and Kathleen Knafl (2005). Compared to other research methodologies, the

integrative review method allows for the combination of diverse methods, such as experimental and nonexperimental research, and has the potential to play a more significant role in evidence-based practice for nursing (Whittemore & Knafl, 2005). Systematic integrative reviews can promote a comprehensive understanding of problems relevant to health care and policy. Data sources accessed during an integrative review can promote a holistic understanding regarding the topic of interest.

The Whittemore and Knafl framework that guided this review consists of five steps: problem identification, literature search, data evaluation, data analysis, and presentation. Through problem identification, a clinical practice issue is brought to the forefront, and the variables of interest are determined. As it relates to the quality care outcomes of patients with SCD, the variables are clinician education, quality care outcomes, and stigmatization. It is important to determine how the variables impact one another to gain further insight regarding the investigative nature of the study. The purpose of the review and variables of interest must be clearly specified in order to better facilitate all other stages of the review and avoidance of complexity (Whittemore & Knafl, 2005).

The next step of the framework is the literature search. The search of literature during this process should be well coordinated and defined to elicit relevant and crucial information. There are numerous databases from which one can generate data, such as, but not limited to, the Cumulative Index to Nursing and Allied Health Literature (CINAHL), Medical Literature On-Line (MEDLINE), and the Agency for Health Care Research and Quality Patient Safety Network. One must be mindful of search terms used to reduce ambiguity with findings. Other recommended approaches of this step are ancestry searching, networking, journal hand searching, and searching via registries (Whittemore & Knafl, 2005). Inclusion and exclusion

criteria should be emphasized at this stage as well for the determination of beneficial primary resources.

The third phase of the framework is data evaluation, where the quality of the primary resources will be evaluated. Evaluating the quality of resources can be complex, as there is no gold standard to make the process more feasible (Whittemore & Knafl, 2005). In assessing the levels of evidence, Melnyk and Fineout-Overholt's (2019) model is a useful tool. Through evidence leveling, one can determine the validity of the research findings.

During data analysis, information will be ordered, categorized, coded, and summarized, through which a consensus conclusion of the evidence can be drawn. Goals of this stage are to reach a comprehensive unbiased interpretation of primary sources and to synthesize the evidence (Whittemore & Knafl, 2005). The risk of error is evident during the data analysis if it is not done systematically. There are five phases of data analysis: data reduction, data display, data comparison, conclusion drawing, and verification. Data reduction involves the determination of an overall classification system in managing diverse methodology data. Data reduction also entails extracting and coding data from primary sources to simplify and organize data into a framework that is more manageable (Whittemore & Knafl, 2005). Data display merely involves the presentation of extracted data in the form of graphs, matrices, charts, or networks that allow for comparison of the primary sources. Data comparison is the process by which themes or relationships of data are examined. The final phases of data analysis are conclusion drawing and verification, in which a higher level of abstraction regarding the data are generated. Similarities and differences are identified; patterns and processes are isolated (Whittemore & Knafl, 2005). With completion of each subgroup analysis, further synthesis of the important elements of each subgroup is conducted relating to the phenomenon of interest. Finally, the review process is

completed and a new conceptualization of the primary sources integrating all subgroups is achieved.

The fifth and final stage of the framework is the presentation. Here, the findings are disseminated or displayed. The conclusion of the integrative review can be reported in a table or in diagrammatic form. To expose the degrees of evidence, keen details from primary resources should be provided. The implications of practice and limitations encountered should also be explained during this stage of the framework (Whittemore & Knafl, 2005). At the end of it all, success of the integrative review is accomplished if there is sufficient valid evidence to influence a quality improvement change.

SECTION TWO: COMPREHENSIVE SYSTEMATIC SEARCH

The goal of the review of literature and data collection is to gain a thorough understanding of the existing research pertaining to the subject matter (Toronto & Remington, 2020). An exhaustive review of literature that met the inclusion criteria was conducted. Databases utilized, as previously aforementioned, were CINAHL, MEDLINE, and PubMed. Search terms used included *sickle cell disease, clinician bias with sickle cell disease, sickle cell disease clinician education, nurse care perceptions and sickle cell disease, sickle cell education, quality care outcomes for sickle cell disease, barriers to sickle cell disease care outcomes, economic burden of sickle cell disease care, stigmatization of sickle cell disease,* and *improving sickle cell disease care.* Utilizing these search terms yielded varying results from the databases. To further expand results, some of the terms were combined using Boolean operators such as *AND, OR,* and *NOT.* Further details regarding the quantification of yielded articles per database will be further discussed.

Terminology

For the research process, it is vital to have a keen understanding of and familiarity with the available databases and terminology that can yield effective results. With aims to achieve a comprehensive and rigorous review, varying databases and other supplemental sources were searched. Platforms through which CINAHL, MEDLINE and PubMed literature were accessed were EBSCO, Ovid, and ProQuest, respectively. Based on the terms used, the databases produced some of the same material. Terms representing the main concepts of the subject matter were carefully selected to elicit a broader representation of results.

Supplemental Search Methods

There were platforms other than the three primary databases from which relevant data were obtained. Websites of organizations such as the CDC, World Health Organization, American Academy of Family Physicians, AHRQ, American Nurses Association, Department of Health and Human Services, and the CMS were searched. Information gathered from the mentioned sites further validated some of the data generated from the literary search. Through the process of hand searching, the American Society of Hematology's *Blood Journal* was explored for articles that were five years old or less and had relevance to the topic of interest. Among 12 related articles located, three met the inclusion criteria.

Definition of Terms

Clinician targeted education—Educational activities that serve to increase or maintain knowledge and professional development of clinicians (Joseph & Huber, 2015).

Continuity of care—Process by which the patient and health care team collaborate for ongoing health care management toward a shared goal of quality care (American Academy of Family Physicians, n.d.).

Equitable care—Provision of care that does not vary in quality based upon one's characteristics, such as ethnicity, gender, socioeconomic status, and geographical location (AHRQ, n.d.b.).

Health literacy—The degree to which an individual has the capacity to obtain, communicate, process, and understand health information in order to make appropriate health information (CDC, n.d.).

Patient-centered care—Care focusing on the patient's particular health care needs (AHRQ, n.d.a.).

Quality care improvement—Actions that lead to measurable improvement in health care services and the well-being of targeted patient groups (AHRQ, n.d.b.).

Stigmatization in healthcare—The act of treating someone unfairly based on character or state of health, which can create barriers to equitable health care (Rao et al., 2019).

Findings

A search of CINAHL using the search term *barriers to sickle cell disease care outcomes* resulted in the identification of five article, two of which met the inclusion criteria. The term *sickle cell disease* was broad, and a search using this term produced over 20 articles. Twelve of the articles were further reviewed for pertinent content regarding the disease state, of which eight met the criteria. The terms *nurse care perceptions* and *sickle cell disease* yielded five articles; however, upon review, none met the criteria. Using the term *sickle cell disease clinician education* produced eight relevant articles, of which five met the inclusion criteria.

A search of the MEDLINE database using the term *economic burden of sickle cell care* returned five articles. Two of the five articles were further reviewed; these met the inclusion criteria. A search using the term *stigmatization of sickle cell disease* produced 12 articles, five of

which met the inclusion criteria. A search of the term *improving sickle cell care* yielded 30 articles, of which 10 were reviewed further, and of the 10, five met the inclusion criteria. Using some of the same search terms previously used with CINAHL produced a significant amount of duplicated results.

Searches through PubMed also produced many duplications from the other aforementioned databases. The term *stigmatization of sickle cell disease* produced four additional sources, of which two were met the inclusion criteria. Over 40 articles were returned from a search of the term *sickle cell disease,* of which eight were reviewed further for relevant information. Most of the information was redundant with prior studies that met the inclusion criteria; therefore, none of these additional sources were included. A search of the term *sickle cell disease clinician education* from this platform led to three additional articles that fit the criteria. An additional search was conducted using the term *nurse care perceptions and sickle cell disease,* and of the six articles produced, one met inclusion. Most of the articles that were excluded from the databases used were studies that focused solely on clinician subjective reports regarding care outcomes of the sickle cell client or the pediatric population and articles that lacked substantial generalizability.

SECTION THREE: MANAGING THE COLLECTED DATA

Following the collection of relevant data, the next phase of the integrative review is to simplify or deduce information to minimize redundancy and achieve high validity to promote a practice change. The recommended methodology from Whittemore and Knafl (2005) for the process of evaluating data was conducted. According to Whittemore and Knafl (2005), during the research analysis process, primary sources should be ordered, coded, categorized, and summarized into an integrated conclusion pertaining to the research issue at hand. During this

process, the aim is to analyze the data and display them, providing a comparison of information gathered, and formulize a conclusion. The data analysis subprocesses to be explored are data display, data comparison, data reduction, conclusion drawing, and verification.

Data Display

To present a visualization of information generated, the data were organized in a chart. Through chart presentation, one can easily gain comprehendible insight on the collected information and interpret themes and relationships. The goal of the visual display, which is in a matrix form, is to aid in presenting the inferences and conclusions that will support the overall dissemination of the study.

Data Comparison and Reduction

With the aid of the visual display, data were assessed for differences and similarities between studies. As per Whittemore and Knafl (2005), relationships, patterns, and themes should be readily identified, as they substantiate the generalizability of the findings. Once patterns can be discerned, the conceptual map can be drawn (Whittemore & Knafl, 2005). Content analysis of the extracted information decreases variability and build on the logical chain of evidence. Additional analysis of data through the comparison process allows for further reduction of redundant information. Comparison of data is imperative, as it aids in identifying gaps or discrepancies amongst studies that could impact validity.

Conclusion Drawing with Verification

In avoiding premature analytic closure of findings or exclusion of pertinent evidence, the investigator needs to thoroughly examine results to a higher level of abstraction (Whittemore & Knafl, 2005). All patterns, themes, relationships, and conclusions require verification with primary resources to establish confirmability and accuracy. Establishing accuracy from the

literature can be fraught with challenges, as results may be equally compelling from high-quality reports (Whittemore & Knafl, 2005). Through all phases of the data analysis process, the goal is to establish an integrated summation of the phenomenon of interest with subsequent promotion of best practice outcomes.

SECTION FOUR: RESEARCH QUALITY APPRAISAL

Quality research should encompass all components of the scientific process. The presented evidence should be robust, ethical, and able to withstand professional scrutiny. For the study at hand, quality was assessed in terms of internal and external validity, as well as the extent to which the design minimized bias or errors. To guide the process of determining the fit of the studies located during the search process, A Measurement Tool to Assess Systematic Reviews (AMSTAR) was used, as this platform can allow for effective interpretation of secondary reviewed sources and diverse study designs (Jamshidi et al., 2018). In assessing the methodological quality of the peer-reviewed studies, 11 domains are evaluated (see Appendix B). Each domain's score ranges from 1 to 5 based upon the criteria met, and the total score is the summation of all 11 domains. A higher score indicates greater validity and quality of research. Though the quality assessment tool can aid in determining research quality, there are some limitations. The tool was not originally devised for quantitative scoring until later revisions; however, the weighting of scores has been debatable, leading to some concerns of reliability (Jamshidi et al., 2018). Despite these concerns, the use of the tool for the studies at hand produced favorable outcomes that were reproducible and highlighted high-quality studies.

Sources of Bias

During the research process, curtailing or identifying systematic errors is imperative. Bias can occur at varying phases of research, which can include the study design, data collection, and

publication (Toronto & Remington, 2020). Common presentations of bias in clinical research include selection, attrition, interviewer bias, performance, and measurement bias. In published studies, there is a high probability that there will be some degree of bias; however, one must consider how the bias can impact the study's conclusions and effective means of avoiding adverse outcomes.

Sources of bias were evident in the reviewed studies. Jenerette et al. (2016), in their mission to evaluate the impact of sickle cell clinician education on the care outcomes of sickle cell patients, utilized majority nurses as participants amongst a cohort of multidisciplinary contenders. Though participation was voluntary, there is some threat to validity of findings considering the participants' varying engagement with patients based upon their roles and responsibilities. Many of the studies evaluated results by conducting pre- and posttests following intervening measures with an educational platform. The goal was to assess what was learned or to reevaluate perceptions of caring for the sickle cell client. Participants of a study who have familiarity with the subject matter can plays a role in selection bias. Studies that conduct random sampling are more likely to minimize selection bias (Jager et al., 2020).

Other evident biases noted in the reviewed studies were attrition and interviewer bias. During the process of several of the investigations, some participants were lost to follow-up due to conflicting obligations. As previously mentioned, most studies utilized pre- and posttests as means of evaluating results of the intervention. Some participants either failed to retest or return to conclude the investigation; Cramer-Bour et al. (2020) discussed this phenomenon during their effort to develop a simulation-based education curriculum.

Interviewer bias was also present in a study spearheaded by Lovett et al. (2017). The focus was on gathering information from providers of an emergency department regarding their

perceptions on caring for patients with SCD. In some cases, it was evident that the interviewer's viewpoints likely interfered with the objectivity of responses. As the interviewee can probably determine what a favorable response should be, it is challenging to validate credibility. In efforts to minimize interviewer bias, questions should be asked as they have been constructed originally, and questions should not be interpreted for the interviewee. Researchers should avoid communicating their personal opinions and maintain a keen awareness of their body language and facial expressions when interviewing participants (Jager et al., 2020).

Internal Validity

Internal validity relates to the accuracy or reliability that studies evoke. Internal validity heightens the reviewer's and the author's confidence that findings are sound with minimal bias. The contents of each study were appraised for quality, validity, and rigor. The various studies evaluated produced different levels of internal validity in that some were consistent with a higher degree of generalizability. Assessing the quality of each study provides a vantage point for determining internal validity. The research process should outline methods to evaluate the quality of the research, as the presence of these methods can aid in determining how closely results of studies approximate to the truth. Through the use of appropriate scientific methods, research can gain means of acquiring increased internal validity. Related literature on the subject matter went through a critical appraisal to determine the studies' quality and, subsequently, their validity.

Appraisal Tools

In addition to the use of the AMSTAR tool to assess the quality of the related literature, a literary matrix was formed to allow for further synthesis of the articles. Review of the generated data can be complex. Thus, the researcher should utilize an appraisal tool that is applicable, and

that can readily promote the identification of information that is valid and reliable. Through rigorous critical appraisal, the strengths, weaknesses, and limitations of the varying studies were explored based upon the AMSTAR checklist and the levels of evidence as adopted from Melnyk and Fineout-Overholt (2019). Other appraisal tools considered for utilization were the Critical Appraisal Skills Program and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA); however, AMSTAR had components that allowed for additional deciphering of the evidence. For the literature matrix and an analysis of literature based on AMSTAR, see Appendices A and C.

Applicability of Results

Study results are congruent in that there is evident benefit of the promotion of continued clinician education on SCD into practice. Barriers exist in practice that can negatively impact the care delivered to the sickle cell client. One's perceptions and suboptimal preparedness to care for patients with SCD were repeated factors that posed impediments to quality care. According to a study conducted by Freiermuth et al. (2016), evident uneasiness with care and bias toward the patient population were significantly decreased with the implementation of continuing education opportunities for providers at the involved health care settings. Efforts to mitigate barriers to the care of the patient population of interest should be ongoing in practice. To generate improved care outcomes for the sickle cell client, there is a definitive need to institute continued clinician education.

Whiteman et al. (2015) highlighted the ill-preparedness of some primary care providers to treat patients with SCD. Other than training received during medical school and residency, there was no specified dedication to the care of this patient population unless one ventured into specialization from a hematological standpoint. In addition, based upon their practice

geographical locations, some providers lack exposure to the care of these individuals, leading to the increased risk of negative misconceptions and mismanagement when opportunities to care for the patient population encounter arise. Yacoub et al. (2019) also shed light on the fact that there is a need for nursing programs to institute more focus on the care and management of patients with SCD so that new nurses can provide appropriate care and obtain a proper comfort level in caring for this patient population when entering the field of nursing. Lack of preparedness and knowledge relating to the disease manifestations, target population potential social dynamics, and treatment options have been shown to heighten the burden of disease and subsequently reduce best practice results (Yacoub et al., 2019).

The findings of the studies are applicable for any practice setting in which there will be potential exposure to the population of interest. Though some health care providers and institutions may have fewer encounters with the patient population of interest due to their demographic region, there remains justifiable cause to incorporate sickle cell management education into their continued education efforts. Being prepared for the inevitable in practice will allow providers to deliver the best possible care for varying health states or conditions.

When one considers the applicability of an initiative, generalizability must also be considered. As alluded to, clinician-based sickle cell education is imperative as well as applicable to all health care settings and therefore, transferability of the benefit to other practice arenas can readily be achieved. Before implementing an initiative, however, one must consider the possible internal and external threats as well as barriers that can impede application of the change. Such barriers may include, but are not limited to, lack of organizational resources, low support from organizational leaders or stakeholders, low support from staff, poor organizational culture, time constraints, and lack of manpower (Murphy et al., 2018).

SECTION FIVE: DATA ANALYSIS AND SYNTHESIS

Comparison

In a review of the literature, there were some notable congruencies as well as some variations. Whittemore and Knafl (2005) contended that patterns, themes, and relationships within the data should be identified, as this will further promote substantiation of the findings. The consensus, based on information gathered, is that continued sickle cell education geared toward clinicians can positively influence clinicians' perspectives on the patient population of interest and care approaches. Methodologies for collecting data and areas of focus differed amongst the studies. Studies were reviewed for similar information, methodologies, and findings, and data were explored based on these patterns.

To acquire information related to the subject matter, some researchers conducted a pre- and post-test analysis after participants were exposed to interventional education. Studies that utilized a pre- and post-test approach with the aim of gaining conclusive findings were those of Bernier et al. (2018), Brennan-Cook et al. (2018), Cramer-Bour et al. (2020), Haywood, Lanzkron, et al. (2015), Haywood, Williams-Reade, et al. (2015), Jenerette et al. (2016), Kayle et al. (2016), Singh et al. (2016), and Yacoub et al. (2019). These studies collectively noted a significant difference in post-intervention test scores, which signified knowledge gains or improved perspectives. Statistical analysis was done within the studies to establish credibility. Some tests were done to assess clinician baseline knowledge regarding SCD management, whereas others focused on clinician perspectives on the sickle cell client and impacts of care.

The remaining studies had varying approaches to gathering information. Surveys and interviews were conducted related to clinician level of comfort with sickle cell management, and perspectives regarding the patient population were examined. Expert opinions of the subject

matter were also analyzed, which provided additional substantiation of repeated congruencies. Oyedeji and Strouse (2020) as well as Matthie and Jenerette (2015), from an expert stance, discussed and provided relative supporting data highlighting the benefit of continued clinician-targeted education to improve care outcomes as well as reduce bias toward patients with SCD.

Minor gaps and discrepancies were identified with the compared research articles. Variances were also noted with demographic regions, as inclusive studies were primarily conducted in North America. Age was also a factor, considering older adults and younger adults. Some of the studies focused on related biases with other disease states; however, they showed relatability to the topic of interest and thus contributed beneficial insight. Additionally, there were different themes and patterns amongst the studies. Identified themes and patterns of the studies will be further discussed in detail.

Thematic Analysis

To achieve further understanding and support of the identified recommended practice change, identified themes and patterns were explored. Through thematic analysis, one can gain grounded meaning from the data that can shed further insight regarding the concept or concepts. Recurring themes identified were brought to the forefront and promoted awareness of needed areas of improvement. Addressing these deficiencies in practice could minimize the negative care experience and the suboptimal quality care outcomes for the sickle cell client. Themes identified include: (a) impact of sickle cell education on care outcomes, (b) barriers to effective SCD management, and (c) patient perspectives regarding care received in clinical practice.

Impact of Sickle Cell Education on Care Outcomes

The burden of SCD is individualized; it varies from person to person. Considering all the potential comorbidities that can be associated with SCD, care often needs to be individualized.

Providers in different specialty areas can significantly contribute to best practice outcomes and effective coordination of care if they are prepared to address the needs of patients with SCD. Providers should have a thorough understanding of the disease process and treatment methodologies. Though education and training related to SCD were likely provided through institutionalized education at various schools, some providers have lacked exposure to the patient population of interest and thus are often ill-prepared to care for these patients effectively once encountered (Haywood, Lanzkron, et al., 2015). To improve care practices for those with SCD, health organizations can benefit from instituting continued sickle cell education for clinicians.

Baseline Knowledge of Providers. The majority of studies in alignment with this theme conducted a pre- and post-test analysis following the implementation of an educational platform that was centered on enhancing clinician SCD management knowledge. Existing knowledge was compared to acquired knowledge following the intervening measure. Yacoub et al. (2019) conducted a study with a control group design involving 77 registered nurses who worked in hematology and genetic units. Pretest and posttest evaluations were conducted to analyze the difference in nurses' knowledge and care practices after the institution of an educational program. The majority of nurses acknowledged that they had discomfort in managing crisis events and pain control for patients with SCD. The study reveals that there is a recognizable need to enhance nursing curricula to bring more focus to SCD management, as this will better prepare them to deliver quality care to this patient population (Yacoub et al., 2019). Findings reflected a statistically significant effect on knowledge scores with a p value of < 0.001, indicating an increase in mean scores for correct best practices following the educational program.

Cramer-Bour et al. (2020), in their comparative pretest and posttest evaluations, examined the care practice knowledge of medical residents before and after they received

simulation-based education on SCD management, particularly in complicated events. Thirty-six medical residents were involved in the study. Results reflected a twofold increase in provider knowledge and correct treatment decision-making following the implemented educational platform, which was sustainable over a 30-day period. Thus, the conclusive stance of Cramer-Bour et al. (2020) is that clinical practice education on SCD will improve patient care and clinician early recognition of potential unwanted events.

Bernier et al. (2018), in their examination of clinician baseline knowledge, focused on clinicians' preparedness to address those in active vaso-occlusive crisis. Participants were exposed to an educational intervention related to SCD manifestations, treatment, and pain management. Pre- and post-surveys were distributed to clinical staff that evaluated overall SCD care knowledge and pain assessments. Results revealed the benefit of enhancing clinician knowledge in SCD management, as there were evident gaps in effective care approaches, particularly relating to pain management. In a similar correlational study, Kayle et al. (2016) conducted an examination of provider gains of knowledge following the implementation of a web-based SCD educational program to promote improved disease management preparedness and quality care. Comparative pre- and post-test evaluations demonstrated clinical knowledge improvements following completion of the educational program for providers, yielding a p value of < 0.0001. Most participants indicated following completion of the program that they gained knowledge that would allow them to be more prepared to care for patients with SCD and the associated complexities.

Ross et al. (2021) took an alternative stance relating to the evident need to improve clinical preparedness to care for patients with SCD. Their study emphasized that it was equally important to address the educational needs of the patient impacted by the disease as a means to

promote partnership in care and quality outcomes. The study, through a purposive sampling of patients and providers from varying health organizations and conferences, examined providers' and patients' understanding of SCD. Findings showed that educational needs exist for both the clinician and the patient that could potentially contribute to adverse outcomes of care. Some providers lacked knowledge regarding new line therapies, acute signs of vaso-occlusion, and pain management methodology for this patient population. Some patients were amiss as it related to triggers that could exacerbate pain crisis events, lacked knowledge of their genetic transferability risk with family planning, and lacked the ability to provide a rationale for their current pharmaceutical therapy for their disease state.

Improving Patient Satisfaction. The lack of quality care for the patient population of interest can negatively impacted patient satisfaction scores for varying institutions. Clinicians who are prepared to take on the complexities of the management of SCD will be better able to meet physical and psychological needs. Freiermuth et al. (2016) conducted a cohort study in which the impact of an ongoing SCD clinician education initiative was examined. Participants, who were selected using convenience sampling, included physicians, nurse practitioners, physician assistants, and registered nurses. Areas of focus were clinician SCD management knowledge and clinician perceptions of the patient population. Surveys were conducted three times over a two-and-a-half-year period. At the end of the study, there were notable positive changes in attitude toward the care of the SCD client and improved care approaches. In addition, patient satisfaction scores showed recognizable improvement from the two participating health organizations.

Singh et al. (2016) focused on examining provider bias and attitudes toward sickle cell patients in need of care, relating mainly to pain management. Participant providers of the study

viewed a video depicting the challenges of patients living with SCD. In addition, pre-test and post-test surveys were conducted to examine the clinicians' perceptions. The implementation of the video was shown to improve the empathetic attitude as well as care practices of the providers involved. Within a 12- to 18-month span, there were improved methods of coordinated care, more positive health outcomes, and increased patient satisfaction scores from those impacted by SCD. Scores reflected an improvement from baseline with a significant p value of < 0.05.

Hospital Readmission. The health care system is subject to a significant financial burden due to the staggering rates of hospital readmission. The 30-day readmission rate for patients experiencing sickle cell crisis events is high following hospitalization (Kumar et al., 2020). According to the 2016 Nationwide Readmission Database, of 67,887 discharges after index hospitalizations, 18,099 (26.9%) patients were readmitted within 30 days of discharge. Amongst the readmissions, 5,166 patients were readmitted within seven days. Sickle cell crisis–associated readmissions in 2016 resulted in a total cost of $609 million, with a total cost of $152 million in the United States (Kumar et al., 2020). In an analysis of the causative variables for the surmounting rates of SCD state readmissions, inadequate management from clinicians was labeled as an associated factor. According to Kumar et al. (2020), understanding the diagnoses and timing of readmissions is imperative to formulate interventions that can reduce readmissions and repair the economic burden. Attempts to curtail the readmission rates of those affected with SCD are critical both from an economic as well as a quality outcome standpoint. The Medicare Payment Advisory Commission suggests that through reduction of hospital readmissions, Medicare could save $12 billion per year (Kumar et al., 2020).

Freiermuth et al. (2016) found that as a result of their two-and-a-half-year cohort study, which focused on improving provider attitude toward caring for patients with SCD and

improvement of patient satisfaction scores, there was an overall decrease in the rate of readmissions. The study's emergency department providers were exposed to an ongoing SCD education initiative. The initiative subsequently allowed for a better understanding in crisis treatment and overall improved clinician attitude in the care of the clients with SCD. Likewise, Brennan-Cook et al. (2018) illuminated in their study that clinician knowledge deficits in managing SCD exist that can negatively impact care outcomes and contribute to frequent hospitalizations and readmissions. There remains a need to prepare clinicians to improve their care practices to care for the patient population of interest effectively. Through education and effective care coordination, institutions can prevent hospital readmissions, reduce the utilization of health care, and contribute to improved quality outcomes (Brennan-Cook et al., 2018).

Barriers to Effective SCD Management

Barriers can be present in patient care that can be detrimental to care outcomes and the overall care experience. These barriers can vary and usually lead to ineffective communication between the medical professional and the patient (Oyedeji & Strouse, 2020). Effects of these barriers include, but are not limited to, decreased patient satisfaction, safety, job satisfaction, and quality care (Brennan-Cook et al., 2018). Compared to other disease states, SCD has been associated with more practice barriers that can interfere with effective care deliverance. To overcome this deficiency in care, it is essential for health care providers to readily identify barriers that could negatively impact care. Nurse leaders have the responsibility to ensure that barriers in practice are being addressed in alignment with patient advocacy. Some of the noted barriers related to SCD care will be further explained. In addition, clinician-targeted education on SCD should address the associated care barriers that could adversely impact best practice.

Clinician Perspectives and Bias. Implicit bias toward patients with SCD exists in practice that hinders the quality of care for those impacted. Several factors have contributed to the emerging bias, including, but not limited to, inadequate provider training, racism, and the opioid misuse epidemic (Bulgin et al., 2018). Studies show that few physicians and auxiliary providers feel confident in treating SCD due to the disease state's complexity (Singh et al., 2016). Provider negative attitudes and bias are significant barriers to optimal pain management (Singh et al., 2016). In practice, there is a need for further education relating to the disease state manifestations, burden of disease, and risk of vital organ damage. In their prospective cohort study, Singh et al. (2016) conducted a comparative analysis on providers' attitudes following provider exposure to a video that depicted patient challenges with pain management and ongoing misconceptions they experience when seeking care. Following the implemented video, a survey revealed notable improvements in provider attitude and perceptions.

According to Kanter et al. (2020), negative clinician perceptions have been shown to deter patients from seeking care and thus subsequently add to the disease cost burden due to lack of management. Matthie and Jenerette (2015) concurred that clinician knowledge gaps regarding SCD contribute significantly to the presenting bias that exists in practice. To mitigate the negative consequences of the perceived biases toward the management of the sickle cell client, education has been deemed paramount, and nurses can be a significant contributor to best practice outcomes (Matthie & Jenerette, 2015). Lovett et al. (2017) concluded from their study that underlying cognitive biases have resulted in misdirected care for those with SCD. Based upon results of clinician interviews, they noted negative perceptions regarding patients with SCD that played a role in suboptimal care approaches, particularly as it relates to addressing pain. The survey demonstrated the need for further provider education on SCD management.

Jenerette et al. (2016) conducted a correlation study to examine the effect of a clinician-focused sickle cell education conference on knowledge and attitude toward patients with SCD. Pre- and post-tests were given for comparison that addressed perceived attitudes in the care of the patients of interest and general care practices. Posttest results showed significant improvement in knowledge scores amongst those who participated in the study. Also noted from the reassessment surveys was a decline in negative attitudes relating to the care approach of patients with SCD. Clinician attendance at the sickle cell education conference positively influenced the participants' overall insight regarding care practices and can validate the benefit of instituting clinician education on the subject matter. Haywood, Lanzkron, et al. (2015) also focused reducing negative attitudes during relative patient encounters. Clinician participants watched a video that discussed SCD and documentary footage on varying individuals' lived experiences with the disease. It was evident from the conducted surveys that many providers lacked understanding of and consideration for the disease burden, which was negatively reflected in their treatment decision-making and the patient-provider relationship. Incorporating the patient's perspective into an educational program will be beneficial an an effort to improve the care experience.

Culturally Competent Care. Cultural competence fosters the acceptance of cultural differences, ethnicities, differences in appearance, customs, and certain rituals. In practice, adhering to principles of diversity and fostering ethical standards will support the deliverance of culturally competent care. Nurse theorist Madeleine Lininger (2001), in her culture care theory, highlighted the importance of cultural competence in practice in promoting the patient's physical and mental well-being. Achieving cultural competence entails continuous development in which

one must have humility, be committed to ongoing professional education to maintain proficiency, and have a keen awareness of self.

According to Williams and Smith-Whitley (2016), providers who aim to understand the lived experiences of those most affected by SCD will inevitably contribute to effective care results. Clinician sickle cell education must be able to enhance cultural competence and foster more equitable care, as this will improve quality care, promote effective treatment management in practice, and improve patient satisfaction (Williams & Smith-Whitley, 2016). Though opposing views may exist amongst the races, contributing to the misconceptions as well as biases toward patients with SCD is the impact of systemic racism as a causative factor. Bulgin et al. (2018) suggested that a lack of communication and understanding between varying groups has contributed to the negative encounters in practice and the development of bias. According to Bulgin et al. (2018), cultural competence training is needed amongst clinicians to enhance diversity and equitable care.

Fragmented Care and Guidelines. Guidelines and quality indicators for the adult client with SCD are few and fragmented compared to those for the pediatric population. Lack of clinical consensus and understanding regarding appropriate treatments for the adult sickle cell population has led to poor care outcomes. Guidance in the treatment of adults often relies on clinical experience or has been extrapolated from pediatric populations due to a lack of research-driven data specific to adults (Adams-Graves & Bronte-Jordan, 2016). While the mortality rate for children has shown a decline, the mortality rate for adults showed an annual increase of 1% from 1979 to 2005 and continues to climb (Adams-Graves & Bronte-Jordan, 2016). Ongoing efforts to improve care and management for the adult patient with SCD is pivotal.

The young adult with SCD transitioning to adult care is often negatively impacted by the burdens of fragmented care. Due to the complexities of the disease, the transition from pediatric to adult medical care can be a high-risk period, as exacerbations of crisis events and complications can arise due to the mismanagement of care approaches. Morbidity and mortality rates for patients with SCD in the United States markedly increases after the age of 18 years (Treadwell et al., 2016). Pitfalls of the transition are due to the failures of the health care providers and the overall health care system in assuring continuity of care. These failures have contributed to significant health care costs that can stifle the economy.

According to Adams-Graves and Bronte-Jordan (2016), improved SCD care outcomes require effective care coordination between hospitals, primary care providers, emergency medicine, and other medical specialists. Clinicians who lack clear understanding regarding effective SCD treatment methods and lack the skills to recognize disease state complications can be a detriment to the patient population of interest (Adams-Graves & Bronte-Jordan, 2016). To improve quality outcomes for the adult population with SCD, there is a need for solidified evidence-based guidelines to guide practice. Until the publication of the *Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014* by the National Institute of Health, there were no evidence-based guidelines for SCD (Adams-Graves & Bronte-Jordan, 2016). All preceding guidelines created were not founded on scientific evidence (see Figure 2).

Care of the sickle cell client needs to be driven by solid evidence. Practicing clinicians should readily have available evidence-based guidelines that can guide the decision-making process in treating patients with SCD. As SCD and treatment options can be complex, many clinicians lack complete comprehension regarding appropriate management. Continued clinical education for SCD and management is imperative, as this will contribute to health care

organizations being in alignment with local and national benchmarks. Clinician-targeted education should be based on evidence to enhance clinical expertise, promote best practice, and aid in bridging the gap toward quality care.

Figure 2

Adult Guidelines for the Management of SCD

Guidelines for the Management of Sickle Cell Disease in Adults			
Source	**Year of publication**	**Title/Description**	**Evidence-based**
WHO	2011	Pharmacotherapy of Sickle Cell Disease	No
Sickle Cell Society (UK)	2008	Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK	No
NIH/NHLBI	2002	The Management of Sickle Cell Disease	No
NIH/NHLBI	2014	Evidence-based Management of Sickle Cell Disease	Yes
NIH/NHLBI: National Institutes of Health/National Heart, Lung and Blood Institute; WHO: World Health Organization			

Note. From "Recent Treatment Guidelines for Managing Adult Patients with Sickle Cell Disease: Challenges in Access to Care, Social Issues, and Adherence," by P. Adams-Graves and L. Bronte-Jordan, 2016, *Expert Review of Hematology, 9*(6), p. 542.

Patient Perspectives Regarding Care

Patients with SCD are often subject to skepticism and bias when they seek care, particularly in the emergency room setting. The most common reason for seeking care is the onset of a pain crisis event. The onset of pain can be severe and last for hours, and thus, having means of controlling the pain is imperative. Despite the debilitating impact of the disease, some patients in the clinical setting have experienced negative aspects of care from providers, which has been associated with poor quality outcomes. Kanter et al. (2020) surveyed various emergency room departments to assess the care perceptions of adults with SCD. Participants

ranged in age from 18 to 60, the majority were African American, and the review was conducted over a 12-month span. The survey used was a fixed-format questionnaire called the Adult Sickle Cell Quality of Life Measure Information System. Some of the themes mirrored focus areas of the Consumer Assessment of Healthcare Providers and Systems surveys. Major problematic areas included perceived lack of empathy, poor communication, and suboptimal pain control. Amongst other components of the survey, one's socioeconomic status was included so researchers could determine whether this impacted the quality of care received. Compared to respondents who were currently working, participants who were not working or received disability indicated that they often experienced bias and were 66% less likely to be satisfied with their emergency department care (Kanter et al., 2020). Though most of the perceived negative engagements took place with clinicians, the study emphasized that patients also held negative perceptions of clerks and or receptionists of the department. Oyedeji and Strouse (2020), in their review of barriers that impact the care experience, offered expert advisory in the promotion of best practice results. In addition to the promotional benefit of continued provider education, their focus was on the importance of collaborative care with the patient as an active participant.

The negative interactions and recurring practice bias toward patients with SCD have been found in some cases to deter patients from seeking care expeditiously. Clinicians who care for those with SCD should develop a keen understanding of the disease process and potential ramifications if care is suboptimal. Those in a crisis event can be subject to acute chest syndrome, cerebral vascular accidents, kidney disease, and other serious complications if efforts are not in place to effectively control the hemolysis effect. To generate improved care outcomes, efforts need to be in place to implement further SCD education in practice and mitigate any stigmatization. According to Haywood, Williams-Reade, et al. (2015), clinician race and

discipline had a relational impact on the associated stigmatization of SCD patients in practice. Lack of exposure to varying races and cultures can contribute to perceived bias, and as previously alluded, there is evident benefit to integrating diversity training into an educational forum. Evensen et al. (2016) concurred that lack of understanding among providers, along with the common associated stereotype of sickle cell clients being "drug seekers," has led to a lack of provider understanding and empathy toward those with the disease. According to Evensen et al. (2016), given the significant burden of disease, there should be practice strategies in place to reduce stigma, prevent delays in service, and promote clinician sensitivity. Clinician-targeted SCD education can serve to decrease bias and support the aims of improving the care experience.

Synthesis of Results

Sickle cell education for clinicians in practice has several advantages, as described in the thematic analysis. Though entities of the disease state might have been covered in most preclinical educational programs, there appear to be gaps in knowledge on how to effectively manage this patient population. According to Whiteman et al. (2015), some graduates have noted that they are ill prepared to tackle some of the psychological and physiological challenges of caring for patients with SCD appropriately. There are errors particularly often during the transition of care from pediatric to adulthood, when the peak of the disease burden is most often triggered. The lack of provider knowledge about how to adequately manage SCD justifies the need to incorporate SCD continued education into practice. Attempts to enhance clinicians' knowledge and readily prepare them to care for the patient population of interest were deemed favorable amongst the reviewed studies. Following the implementation of an educational intervention, some studies noted improved patient satisfaction scores and a decrease in hospital

readmissions associated with SCD crisis events. Providers should carry out practice that aligns with the best supportive evidence, as this will promote gains of quality outcomes.

In addition to the pitfalls of lack of clinician preparedness to care for those with SCD, there exist other barriers to effective sickle cell management. As previously stated, the barriers include clinician bias, lack of cultural care competence, fragmented care, and lack of guidelines. These barriers pose threats to the overall quality of care these patients receive. The perception of addiction has been highly associated with the SCD client, leading to the recurrent presentation of implicit bias in practice (Brennan-Cook et al., 2018). Other related studies presented similar findings that the deep-seated stigmatization impedes the ability to provide appropriate evidence-based care for these patients.

Lack of cultural competence was named as a barrier, as being in alignment with this has been found to enhance one's empathetic nature in practice (Williams & Smith-Whitley, 2016). Stereotyping and prejudging lends to being culturally incompetent as this prevents the ability to understand, appreciate and interact with people of differing cultures, practices, or beliefs. Sickle cell disease impacts mainly those of African ancestry followed by Hispanic, Middle Eastern, Asian, and Mediterranean descent (Centers for Disease Control and Prevention, 2019). Considering those individuals that may be impacted, race and exposure also play a role in the care outcomes. To achieve the physiologic and psychological well-being of the patient, methods to improve clinicians' cultural competence should be incorporated into the continued education platform. Lack of understanding and effective communication between the patient and clinician have been associated with adverse outcomes in the case of differing cultures or races (Bulgin et al., 2018).

As another barrier to effective SCD management, the fragmentation of care and treatment guidelines has been disadvantageous to the care approach. When compared to the care of the pediatric population impacted with SCD, evidence-based guidelines for managing SCD in the adult client is lacking. Care of the adult has relied heavily on practice experiences rather than sound evidence. Until the National Institute of Health's formulation of evidence-based guidelines in the management of SCD for the adult client in 2014, no guidelines were evidence-driven (Adams-Graves & Bronte-Jordan, 2016). Lack of concrete guidelines for practice leads to care fragmentation and bears the risk of negative outcomes of care, particularly for those with SCD, considering all of the potential complications one can endure. The fragmented care surrounding SCD is also evident and most common during the transition period when individuals must begin to seek management of their disease as an adult rather than a pediatric patient (Treadwell et al., 2016). Fragmentation of care was a common element in the related studies reviewed, justifying the need for improvement. Unfortunately, the existing inadequacies in preparing the patient for effective transition of care can lead one to seek emergency medical attention. Considering the disease burden for those directly impacted by SCD, as well as the overall financial burden to health care organizations, efforts to promote best practice based on evidence should be implemented. Effective management of the disease state and coordination of care are avenues for quality outcomes. To mitigate suboptimal care decisions, continued clinician-targeted education should incorporate evidence-based guidelines to achieve effective uniformity and coordination of care.

Another repeating theme within the literature was the patients' perspective on the care they receive. The lived experience of those impacted with SCD should be examined to promote the empathetic aspects of care. The vaso-occlusive nature of SCD can trigger debilitating effects,

with pain being the greatest manifestation. Gaining pain control is the most pivotal objective of the patient, which at times may create a negative reaction from the health care provider. As previously mentioned, patients with SCD have been characterized as "drug-seekers" or known in the medical community as "sicklers." Related literature regarding the patient's perspective on the subject matter concurs that the negative character associations create a deterrence for seeking medical care (Jenerette et al., 2016). There were noted references to lack of empathy, lack of active listening to concerns, and ineffective communication styles. These deficiencies of the care experience support the benefit of instituting further education regarding SCD and management in practice. Aims to generate further understanding of the lived experience of individuals impacted by this condition, along with the disease burden, can guide the way toward best practice and quality care.

Descriptive Results

The recurring themes as discussed support the need for the practice initiative. Despite education and training from the varying medical and nursing programs, there is a need to improve clinician care practices in managing those with SCD. Impeding factors that limit quality care should be addressed in practice. In review of the related literature, the importance of clinician sickle cell education, barriers to effective SCD management, and patient perspectives regarding their care encounters were repeated themes that triggered the need for further inquiry. In the studies, there was great focus on emergency care medicine and interactions along with care practices of clinicians of this specialty area. Though the main focus was on emergency care practices, it was evident from Whiteman et al.'s (2015) study that most primary care providers have limited experience in caring for patients with SCD; thus, their lack of prudence can contribute to fragmentation and subsequent poor care coordination. Conclusively, continued

sickle cell clinician education is warranted at many fronts of the medical arena. Clinicians who can benefit include not only those working in emergency medicine, but providers from all specialty areas, as those with SCD may have other comorbidities.

Ethical Considerations

In an integrative review, there are no direct interactions with human subjects. In accordance with rules guarding protected health information, participants from the varying studies were not identifiable, as information such as name, date of birth, address, or admission history was not provided. The project leader ensured that studies reviewed upheld ethical principles such as autonomy, beneficence, justice, and non-maleficence as to be in alignment with moral decency. To further enhance ethical standards of practice and in preparation to conduct the project in accordance with the expectations of Liberty University's Institutional Review Board, the leader as well as the project chair completed training through Collaborative Institutional Training Initiative (see Appendix E). When all requirements were met, approval to proceed with the integrative review was granted by the Institutional Review Board (see Appendix D).

Timeline of Project

Planning for the integrative review began in September 2020, when the project leader initiated gathering information regarding the subject matter. During the planning and data collection stages, the goal was to determine if there was a sufficient amount of available data to answer the clinical inquiry. For two to three months following the systematic search, the data gathered were keenly analyzed for evident validity. Approval to proceed with the project, as previously indicated, was granted in January 2021 by Liberty University's Institutional Review Board, and writing began. *A Step-by-Step Guide to Conducting an Integrative Review* by Toronto

and Remington (2020) was used to direct components of the review. Additional guidance was gained through assistance of the project chair. Overall completion of the project took about six months from start to finish.

SECTION SIX: DISCUSSION

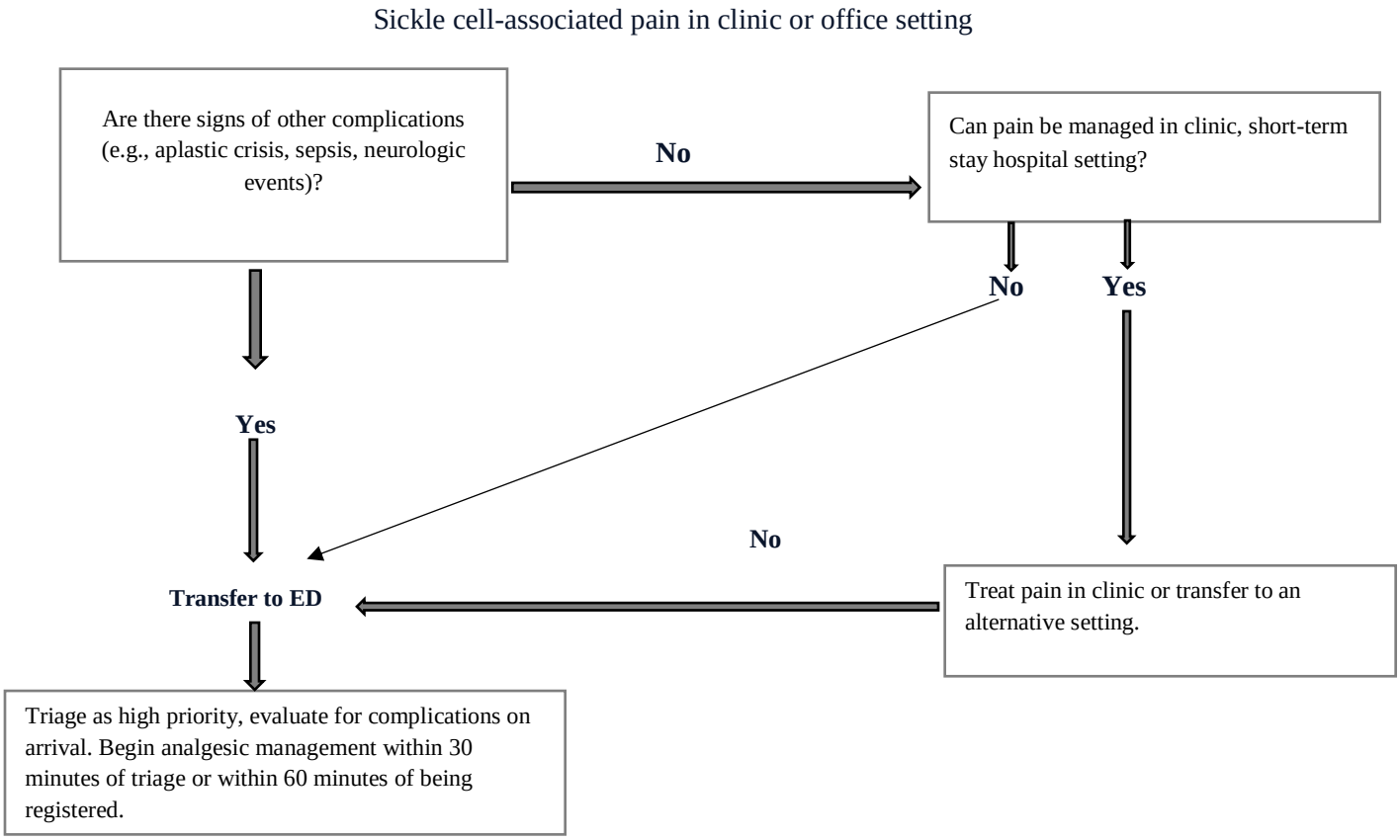
The reviewed literature validates that there exists the need to better prepare health care providers to care for patients with SCD effectively. The obstacles to quality care can be attributed to lack of knowledge relating to the disease burden risk, stereotypes, lack of treatment guidance, health care disparities, and lack of continuity of care. When compared to other common disease states such as heart disease or cancer, SCD remains among those that lack nationwide efforts in health promotion and education. According to Haywood, Williams-Reade, et al. (2015), racial bias has played a role in the disparity of care, and efforts to promote more awareness through national education and health promotion for SCD is needed. Due the risk of adverse outcomes, efforts should be made to mitigate the stereotypes associated with SCD. Clinical practice should be culturally and racially unbiased, as this can promote improvement with the care experience.

Amongst the manifestations associated from this disease state, pain is the most debilitating as the vaso-occlusive process progresses. Once a pain crisis presents, there is great focus on the alleviation of pain from the vantage point of the patient. Unfortunately, the current opioid epidemic has made it challenging for patients with SCD to receive the pain medicine they need and has contributed to the health care provider's skepticism toward the impacted patients. For acute and chronic pain associated with SCD, the mainstay for pain management has been opioid analgesics (Brandow et al., 2020). At the peak of the opioid epidemic, the CDC issued guidelines that deterred the practice of prescribing narcotics for conditions that were deemed

inappropriate. The CDC further clarified, as requested by the American Society of Hematology, the American Society of Clinical Oncology, and the National Heart, Lung, and Blood Institute (NHLBI), that the guidelines did not pertain to patients with cancer or SCD who have acute or chronic pain (Brandow et al., 2020). Considering the opioid crisis and the added benefit of narcotics therapy in controlling the pain of the SCD client, there is justified benefit to promoting clinician education that will also address steps to appropriate pain control and narcotic prescription monitoring. To better support the clinician in addressing pain related to SCD, particularly in acute care settings, guidelines developed by NHLBI can be utilized (see Figure 3).

Figure 3

Algorithm for Acute Pain Management in Patients with SCD



- There should be prompt treatment of pain, first dose of analgesics before transfer if possible or within 30 minutes of arrival; give second dose if delay in transfer.
- Give intravenous or subcutaneous opiods (morphine or hydromorphone) per patient-specific protocol
- Reassess for pain sedation every 15 to 30 minutes; readminister analgesia until pain relief obtained. Consider dose escalation by 25% until pain controlled.
- Use nonpharmacologic approaches such as heat; manage pain every six to eight hours. If unable to control pain, consider admission to short-term observation unit or hospital.
- Begin patient-controlled analgesia in the ED, if possible (or once admitted if not initiated in ED).

Note. Adapted from National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease, 2015.

To achieve quality care outcomes, health care providers must gain full preparedness through education and exposure to be able to manage the complexities of SCD appropriately. All

practice disciplines should integrate a continued education program with focus on this disease state, as all parties will have a part in care. As noted from the literature, health care professionals in practice have felt ill prepared to manage the varying intricacies of SCD. Others lack exposure, which may lead to mismanagement. In conjunction with the aforementioned deficiencies, the existing negative perceptions associated with this patient population has led decreased displays of empathy and ineffective patient-provider relationships in practice. To promote improved care outcomes, health care institutions should adopt programs that can foster continued clinical education on SCD.

Implications for Practice

The findings of the integrative review are key to improving further nursing education and care practices for the sickle cell patient. Study findings bring to light that most providers are not prepared to tackle the complexities of SCD. The existence deep-seated biases in practice toward this patient population that have also contributed to the negative care experience was commonly noted throughout the literature. Nurse educators can explore the results of the review to devise methods to improve their pathophysiology curriculum to cover this material more thoroughly. Additionally, nurse educators can incorporate strategies to clarify some of the common misconceptions about the disease state.

Particularly as it relates to adults with SCD, it has been noted that guidelines directing the course of practice are lacking, leading to the fragmentation of care approaches. Gaps in care contributing to increased acute hospital visits have occurred during the transitional period from pediatric disease management to the adult stage. Failures of the transition have been attributed to the inadequate promotion of the continuity of care and patients being uninsured or underinsured. Through their education and training, the advanced practice nurse leader can spearhead efforts to

improve care across the continuum, develop guidelines, and promote the integration of guidelines into the practice setting. As a role model and patient advocate, the nurse leader should strive to mitigate health care disparities for this patient population. This can facilitate best practice outcomes.

Knowledge deficits relating to care approaches for the sickle cell patient and deep-seated biases have negatively impacted the quality of care. There is great benefit in instituting clinician-targeted sickle cell education in practice that can serve to improve the treatment decision-making process and the provider-patient encounter. Findings of the review promote awareness of the practice pitfalls related to the care of those with SCD and can inspire health institutions and providers to examine their engagement process and care outcomes of this patient population. The advanced practice nurse leader can utilize the findings of the study to examine the knowledge and understanding of SCD of their fellow nurses and auxiliary staff, which can aid in developing a clinical education program.

Application of the AACN DNP Essentials

The Doctor of Nursing Practice (DNP) student must consider applying the American Association of Colleges of Nursing (AACN) Essentials of Doctoral Education for Advanced Nursing Practice Competencies when instituting a practice initiative. The eight components of the DNP essentials are structured to promote the highest level of nursing practice (AACN, 2006). The essence of conducting an integrative review is in alignment with Essential I, involves a critical review of the literature to find evidence-based data to support the project inquiry. Essential II considers the complex needs of humankind. The DNP student is uniquely prepared to contribute to the nursing profession and science by translating and disseminating research into practice (AACN, 2006). Essentially, once there is an identified deficit in care, such as those

discussed relating to SCD, there needs to be the incorporation of evidence-based interventions to improve practice. With Essential III, there is the incorporation of systems thinking and leadership for the facilitation of organization-wide changes (AACN, 2006). Aims of sustainability for the practice change are readily achievable when the organization collectively gains support for the initiative. The DNP-prepared provider in compliance with this essential is tasked with ensuring patient safety, addressing ethical dilemmas, and ensuring that the practice change is based on scientific research. In current practice, the transformation of health care is further enhanced through the varying capabilities of information technology. Compared to the past, patient care today has more readily available data that can significantly impact care outcomes and decision-making efficiencies. Essential IV focuses on the use of technology in practice, which can promote care efficiency, safety, and overall improved patient-centered care (AACN, 2006). Means of retrieving information for the integrative review on the subject matter require the DNP student to acquire skills in computer-based programs and retrieval of data. Information gathered can support the practice change, nurse leadership, quality improvement, and effective treatment decision-making.

Essential V focuses on addressing health care policy for advocacy in health care (AACN, 2006). This study promoted awareness regarding the disparities of care that those with SCD may encounter. The DNP student will contribute to promoting awareness of the identified problem within the health care delivery system through application of Essential V. To improve care approaches, particularly for the underserved, the DNP-prepared nurse can aid in the development of improved health care policies on local, state, or national platforms. As mentioned, those with SCD are often subject to ineffective collaborative care, particularly during the transition of care from pediatrics to adulthood.

Essential VI emphasizes the importance of professional collaboration to improve patient and population health outcomes (AACN, 2006). Continuity of care through effective collaboration will serve to improve the quality of care for those with SCD and decrease the need for recurrent hospitalizations related to vaso-occlusive crises. The DNP student who fulfills this essential will be prepared to effectively lead interprofessional teams, resulting in the improvement of the patient care experience. SCD in the United States has contributed to significant financial burden due to the climbing health care costs. In an effort to curtail both the physical and financial impact of this disease, the DNP leader should coordinate measures to improve health promotion and crisis risk reduction. Essential VII focuses on clinical prevention and population health. Through the promotion of clinician-targeted sickle cell education and patient supportive services, quality of life and care outcomes for the patient population of interest can be improved.

Essential VIII, the final essential, aims to advance nursing practice. The practice issue was identified as one that could cause long-term negative physical and psychological consequences for the patient. The overall goal of this essential is to improve patient outcomes across the board. The DNP leader must promote change at the systems level with the endorsement of an evidence-based care approach. Findings of the study signify the benefit of the implementing the proposed SCD education in practice. The leader must develop skills to motivate gains of support from relative team players. This integrative review can serve as motivation for fellow nurses and educators to develop care approaches that will improve quality care.

Limitations of the Study

Though the studies revealed significant information, there were some evident limitations. Data specific to sickle cell education for practicing clinicians were limited. There was great focus on comparing health care providers' existing knowledge regarding SCD with gained knowledge after an educational intervention; however, there were no transparent recommendations of sustainability. A majority of the studies relied on convenience sampling, which impacted the generalizability of the results. Subjects were pooled from existing work environments and were mainly physicians, nurses, and other advanced practice providers. Based on the projects' nature and the varying practice environments, it was challenging to determine how extensively bias was controlled for amongst the participants.

The health care environment of focus mainly pertained to emergency room settings, which limited the practice inquiry association with other specialty areas. As studies primarily focused on the emergency room setting, this posed a challenge to control for differences in the health care environment that can impact the quality of care. Nevertheless, the health care environment is relevant to the study, as it could substantiate further validation that an education program is beneficial to practice, particularly in the case of those with limited exposure in the care of patients with SCD. Future investigations should include a broader emphasis on specialty areas such as primary care, pain management, hematology, cardiology, and nephrology.

Gaps in the literature exist pertaining to practice barriers and strategies for overcoming them. Relating to the patients care experience, low patient satisfaction scores were factored in and viewed as reasonable cause to institute a practice change. In addition, there was no clear guidance from the related studies to improve these deficiencies. Despite the identified limitations, there was substantial information from the studies that can influence a practice

change and improve care practices for patients with SCD. The research findings and their limitations can inspire clinicians and the nursing profession to formalize methods to improve care practices for those impacted by SCD.

Dissemination

The student will present study findings and the implications for practice to nursing faculty colleagues and students at Liberty University. During the DNP project defense, the aim is to promote an awareness of the practice issue and present a rationale for the quality improvement initiative. The DNP student plans to present study findings to fellow nurses, physicians, advance practice providers, and other auxiliary personnel of the local hematology-oncology practice in which they are presently employed. The DNP student also plans to correspond with the director of an affiliated local emergency room department to address their care practices for the SCD patient population; findings of the study will be shared as means to identify potential ways of instituting clinician sickle cell management training. Support from other health care professionals has been sought to present educational ideas to community primary care practices to improve the continuity of care for patients with SCD. The DNP student has been invited to discuss the practice issue with their local chapter of the Maryland Sickle Cell Disease Association during an upcoming virtual patient appreciation event. Study findings will be made available for the public to encourage continued studies on the matter and motivate continued quality improvement efforts.

Conclusion

SCD is an inherited disease that can have a debilitating effect on those impacted. The most significant physical burden is pain due to the vaso-occlusive nature of this disease state, which can negatively impact vital organs. In the United States, SCD, particularly as it relates to

crisis pain events, is a significant contributor to health care financial burdens and heightened 30-day hospital readmission rates. Despite the associated morbidity from SCD, quality of care for most patients is suboptimal. In clinical practice, biases exist toward this patient population that have negatively impacted the deliverance of care. Additionally, according to the literature, there are practicing clinicians who are ill prepared to address the multivariate complexities of this disease state from their formal education and training. Race, socioeconomic status, and opioid regulations are other associated factors contributing to the deficiencies in the care approach. As a means to improve quality care outcomes for patients with SCD, clinician-targeted SCD education in practice should be instituted as part of the health care organization's competency training. Study findings promote awareness of the practice issue and justify the need to strive toward best practice outcomes. The adopted SCD education program should also address the physiological components of managing the disease state, as well as the factors that can subsequently create bias or stereotypical perceptions. Diversity or cultural competence training would be beneficial to incorporate into the lesson plan.

The DNP student has been prepared through education and training to spearhead efforts to formulate a SCD clinician education program at their current health care institution. Based upon findings in the literature and through promoting awareness of the practice issue, it is hoped that fellow clinicians and leaders will be inspired to incorporate this initiative in their practice. As a nurse leader, the DNP-prepared provider is called to model patient advocacy, particularly in the case of sickle cell patients who may be subject to deficiencies in care.

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Appendix A

Literature Matrix

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Adams-Graves, P., & Bronte-Jordan, L. (2016). Recent treatment guidelines for managing adult patients with sickle cell disease: challenges in access to care, social issues, and adherence. *Expert Review of Hematology*, *9*(6), 541–552. https://doi.org/10.1080/17474086.2016.1180242	To promote awareness that premature mortality occurs for those with sickle cell disease despite advances in research and medicine. In addressing challenges evidence-based guidelines has been developed to guide practice as this has been lacking.	No sampling. Sickle cell care practices were reviewed. Identified gap in transitional care from pediatrics to adult leading to increased disease burden, hospital readmissions and inappropriate utilization of resources.	Qualitative/ Narrative	Level 7	Contributing to poor outcomes of care is the conclusion that a large amount of adult providers lack knowledge on how to care for adults with SCD, leading to poor management and complications. The availability of evidence-based guidelines developed by the NHLBI and CDC will have great potential to improve quality care.	There are valid points presented however the utilization of the new guidelines was not generalized at the time of the expert narrative. Though guidelines may be available to guide practice there is no certainty that they will be used consistently with varying providers.
Bernier, K. M., Strobel, M., & Lucas, R. (2018). Assessing the effect of an educational intervention on nurses' and patient care assistants' comprehension and documentation of functional ability in pediatric patients with sickle cell disease.	To determine the benefit of clinician educational intervention in improving care and documentation patients with vaso-occlusive episodes.	Convenience sampling involving clinician staff (nurses and patient care assistants). Pre/post surveys measuring SCD care knowledge and compliance of pain functional	Descriptive Correlational	Level 4	Improving staff knowledge regarding SCD care improved clinical recognition of benefit and utilization of the acute functionality assessment tool.	Sample size of participants was small and thus limits the studies generalizability and comparability to other settings in the care of those with sickle cell disease.

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Journal of Pediatric Nursing, 41, 117–122. https://doi.org/10.1016/j.pedn.2018.04.001		ability assessments.				
Brennan-Cook, J., Bonnabeau, E., Aponte, R., Augustin, C., & Tanabe, P. (2018). Barriers to care for persons with sickle cell disease: The case manager's opportunity to improve patient outcomes. *Professional Case Management, 23*(4), 213–219. https://doi.org/10.1097/NCM.0000000000000260	To review and identity barriers that can impact care of those with sickle cell disease (SCD).	Convenience sampling involving patients with SCD at emergency departments and the impact of case management promoting improved care outcomes.	Descriptive Correlational	Level 4	Barriers identified and specific interventions by case management was found to contribute to improved outcomes.	Lacks some generalizability as the study was done at a single setting. Though findings are likely valid this could be further confirmed if there were comparative analysis from other facilities or settings.
Bulgin, D., Tanabe, P., & Jenerette, C. (2018). Stigma of sickle cell disease: A systematic review. *Issues in Mental Health Nursing*, *39*(8), 675–686. https://doi.org/10.1080/01612840.2018.1443530	To examine literary findings regarding health-related stigmatization in adolescents and adults living with sickle cell disease.	No sampling. Review of existing literature regarding consequences, the effect of stigma on psychological well-being, effect on physical well-being and patient-provider relationship.	Qualitative	Level 7	Findings collectively reveal that SCD stigmatization exists and can have detrimental consequences. Contributing factors to the stigma were racism, disease status, pain and opioid usage. Proposal need of public and medical effort to decrease the stigma to gain improved care outcomes.	Findings were relative to the reviewed literature on the subject matter posing limits of generalizability.

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Cramer-Bour, C., Peterson, J., Walsh, B., & Klings, E. (2020). Practice makes perfect: Education in sickle cell disease. *CHEST, 158*, A1338-A1339. https://doi.org/10.1016/j.chest.2020.08.1215	To propose development of a simulated-based curriculum that would increase education for intensivists taking care of those with sickle cell disease.	Purposive sampling with medical residents who took pre-post tests before and after an sickle cell education simulation; rating their knowledge of sickle cell management particularly in complicated situations.	Descriptive Correlational	Level 4	Following the curriculum there was a two- fold increase with provider knowledge which was noted to be sustained over a 30 day period. The proposition is that education tactics to this liking will improve patient care.	Limited to focus of medical residents in terms of those receiving the simulated-based education though there was identified discomfort amongst intensivists in managing patients with sickle cell disease.
Evensen, C. T., Treadwell, M. J., Keller, S., Levine, R., Hassell, K. L., Werner, E. M., & Smith, W. R. (2016). Quality of care in sickle cell disease: Cross-sectional study and development of a measure for adults reporting on ambulatory and emergency department care. *Medicine, 95*(35), Article e4528. https://doi.org/10.1097/MD.0000000000004528	To examine outcome measures of the quality of ambulatory and emergency department care for patients with sickle cell disease.	Purposive sample of adult patients with sickle cell disease. Survey given to assess their perceptions of care received from their visits. Study done over a 7 month period.	Cross-sectional	Level 2	Identified themes: Provider lack of understanding of SCD. Perceived disrespectful providers and care bias. Proposal of provider education to improve care and patient satisfaction.	Sickle cell continued education implementation would be beneficial, no education platforms provided or suggested. More efforts of generalizability should be explored.
Freiermuth, C. E., Silva, S., Cline, D. M.,	To assess provider attitudes	Convenient sample of	Longitudinal/ Cohort	Level 4	During the 2.5 year period there was noted	Anonymous surveys and thus uncertain if

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
& Tanabe, P. (2016). Shift in emergency department provider attitudes toward patients with sickle cell disease. *Advanced Emergency Nursing Journal*, *38*(3), 199–212. https://doi.org/10.1097/TME.0000000000000106	and care approaches toward patients with sickle cell disease over a 2.5-year timeframe with the implementation of ongoing SCD education.	providers from 2 teaching institution sites; emergency department. Perception about sickle cell disease patient scale survey given 3 times over a 2.5 year period in assessing perceptions following efforts of continued clinical education.			positive changes in attitude amongst ED providers. Negative attitudes decreased and thus there were notable improved sickle cell patient satisfaction scores, and care approaches.	sample groups involved the same providers. High turnover rate at both institutions with the potential of new staff with different attitude for accurate comparison. Lack of generalizability as sites were from different regions.
Haywood, C., Lanzkron, S., Hughes, M., Brown, R., Saha, S., & Beach, M. C. (2015). The association of clinician characteristics with their attitudes toward patients with sickle cell disease: Secondary analyses of a randomized controlled trial. *Journal of the National Medical Association*, *107*(2), 89–96. https://doi.org/10.1016/	The purpose was to assess the ability of an educational video to improve attitudes that clinicians have toward patients with sickle cell disease. Aim to validate that negative attitudes toward the patient population can create barriers to high quality care.	Random sampling involving clinicians from varying medical wards at a medical institution. Surveys completed in assessing their overall perceptions toward caring for those with SCD prior to watching an educational video.	Randomized control trial	Level 2	Clinician race and discipline played an important part in the exhibited attitudes toward the patient population yielding p-values of <0.001 and 0.004 respectively. There was identified need to incorporate a cultural/racial sensitivity focus into clinician education.	Small sample size and limited generalizability as study was conducted at a single institution.

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
S0027-9684(15)30029-8						
Haywood, C., Williams-Reade, J., Rushton, C., Beach, M. C., & Geller, G. (2015). Improving clinician attitudes of respect and trust for persons with sickle cell disease. *Hospital Pediatrics, 5*(7), 377–384. https://doi.org/10.1542/hpeds.2014-017	To test the effect of high intensity and low intensity educational intervention designed to improve provider attitudes toward youth with sickle cell disease.	Convenience sampling involving patients with SCD at emergency departments and the impact of case management promoting improved care outcomes.	Descriptive Correlational	Level 4	Both interventions tested elicited improvements in the SCD attitudes expressed by providers. Reduced negative attitude score and improved positive attitude scores, yielding P value <. 001. Higher intensity elicited stronger effect, however.	Providers cared for patients mostly of the pediatric population. There is uncertainty of results would be similar for adult patients. There is subject of bias based upon the patient population age.
Jenerette, C. M., Brewer, C. A., Silva, S., & Tanabe, P. (2016). Does attendance at a sickle cell educational conference improve clinician knowledge and attitude toward patients with sickle cell disease? *Pain Management Nursing, 17*(3), 226–234. https://doi.org/10.1016/j.pmn.2016.05.001	To compare clinician SCD knowledge and attitudes towards patients with sickle cell before attending conference and post-conference.	Purposive sampling of providers at a sickle cell educational conference Pre-post tests given in assessing impact on clinician knowledge.	Descriptive Correlational	Level 4	Post conference showed improvement in knowledge scores from those who participated. There was also noted decline in clinician negative attitude in the care approach of those with sickle cell disease.	Sample size of healthcare providers could have been larger in achieving more generalization. With questionnaires being maintained as anonymous it is not possible to track within subjects' responses.
Kanter, J., Gibson, R., Lawrence, R. H., Smeltzer, M. P., Pugh, N. L., Glassberg, J.,	To examine the sickle cell related care experience of adolescence	Purposive sampling of patients with SCD at various Sickle	Descriptive/ Correlational	Level 4	Negative perceptions in practice highly correlated to poor outcomes of care and	Study was limited to 8 practice locations limiting generalization of

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Masese, R. V., King, A. A., Calhoun, C., Hankins, J. S., & Treadwell, M. (2020). Perceptions of U.S. adolescents and adults with sickle cell disease on their quality of care. *JAMA Network Open, 3*(5), Article e206016. https://doi.org/10.1001/jamanetworkopen.2020.6016	and adults impacted with SCD.	Cell Disease Implementation Consortium (SCDIC) sites. Surveys distributed focusing on pain management, care quality, and self-efficacy.			the care experience for those with SCD. Most patients concurred that there was evident lack of empathy in practice.	findings. Respondents were not obligated to answer all questions on survey and thus response rate differed for each variable.
Kayle, M., Brennan-Cook, J., Carter, B. M., Derouin, A. L., Silva, S. G., & Tanabe, P. (2016). Evaluation of a sickle cell disease educational website for emergency providers. *Advanced Emergency Nursing Journal, 38*(2), 123–132. https://doi.org/10.1097/TME.0000000000000099	To examine the effectiveness of website educational modules in improving knowledge among health care providers and nursing students in the care of patients with sickle cell disease.	Convenient sample of ED providers (nurses and physicians) to assess the accuracy and effectiveness of a website educational program for sickle cell disease management. Survey of website along with pre-posttests of modules.	Descriptive Correlational	Level 4	The website was found to be a useful tool in providing education and evidence-based resources to better prepare providers as well as nursing students to care for patients with sickle cell disease. Improved knowledge from the viewing videos.	The sample size was small particularly as it related to the participant providers (nurses and physicians) and therefore was not statically analyzed as with the nursing students which yielded a p value of < 0.0001
Lovett, P. B., Sule, H. P., & Lopez, B. L. (2017). Sickle cell disease in the emergency department.	To examine provider underlying cognitive biases that could	Convenience sample in the selection of caregivers for in-depth interviews to	Qualitative	Level 6	Surveys demonstrated need for further education regarding sickle cell disease care and management.	Findings were similar to other relatable studies conducted however lacked some

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Hematology/Oncology Clinics of North America, 31(6), 1061–1079.	misdirect care of those with sickle cell disease.	learn about their perceptions regarding the care of patients with sickle cell			Common misconceptions seemed to be an interfering phenomenon with the care approach.	generalization as the focus only involved one hospital institution.
Matthie, N., & Jenerette, C. (2015). Sickle cell disease in adult: Developing an appropriate care plan. *Clinical Journal of Oncology Nursing, 19*(5), 562–568. https://doi.org/10.1188/15.CJON.562-567	To examine clinician knowledge gaps in caring for the sickle cell client.	No sampling. Care practices in sickle cell disease management based upon varying cases were reviewed	Qualitative/Narrative	Level 7	Education is imperative in mitigating personal biases toward the management of the sickle cell client. Nurses play a key role in education and advocacy in promoting best practice outcomes.	Clinician education is an identifiable benefit in the care of the sickle cell client however there is no guarantee in lessoning the internal bias associated with this patient population.
Oyedeji, C., & Strouse, J. J. (2020). Improving the quality of care for adolescents and adults with sickle cell disease—It's a long road. *JAMA Network Open, 3*(5), Article e206377. https://doi.org/10.1001/jamanetworkopen.2020.6377	To examine barriers to quality care outcomes of patients with sickle cell disease and strategies for improvement.	No sampling. Strategies to reach high-quality care for patients with SCD in U.S. was examined.	Qualitative/ Narrative	Level 7	Several strategies were proposed to improve care outcomes. Multidiscipline approach, provider/patient education, tele monitoring programs aid aims to improve equitable care are amongst recommended methods to improve care.	There are valid points presented that can heighten care outcomes however there is no definitive means to address funding and resources for the initiative.

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Ross, D., Sinha, C., Bakshi, N., & Krishnamurti, L. (2021). Educational needs of patients and caregivers living with sickle cell disease results in development of web-based patient decision aid. *Journal of Advanced Nursing, 77*(3), 1432–1441.	To determine how patients and providers have received education about sickle cell disease and their understanding and knowledge about available therapies.	Purposive sampling of patients and care providers from varying health care organizations and conferences.	Qualitative/ Descriptive	Level 4	Educational needs for both provider and patients identified justifying need to design educational strategies for both parties in generating improved care outcomes.	Lack of generalizability as participants of study were also obtained from conferences which may input suboptimal representation of the population of interest.
Singh, A., Haywood, C., Beach, M.C., Guidera, M., Lanzkron, S., Valenzuela-Araujo, D., Rothman, R. E., & Dugas, A. F. (2016).Improving emergency providers' attitudes toward sickle cell patients in pain. *Journal of Pain & Symptom Management, 51*(3), 628-632.e3.	Evaluation of provider bias and negative attitudes causing barriers in the management of pain for those with sickle cell disease.	Convenience sampling of providers from an emergency department exposed to viewing video depicting patient challenges living with sickle cell disease. Providers were given pre and post surveys evaluation their perceptions.	Descriptive/ Correlational	Level 4	Video-based educational intervention showed to improve emergency providers' attitude toward patients in pain with a sickle cell crisis event. There was notable improved health outcomes and increased patient satisfaction scores within 12-18 months. Scores showed improvement from baseline showing P value of < 0.05.	The intervention was administered at one academic institution where patients and providers in the video were from the same institution. Results of the study can be prone to selection bias as participant providers may be more apt to changing their attitudes versus those who did not participate.
Treadwell, M., Johnson, S., Sisler, I., Bisko, M., Gildengorin, G., Medina, R., Barreda, F., Major, K.,	To evaluate hypothesis which states that ratings of self-efficacy positively	Purposive sample of patient participants from two sickle cell focused institution	Descriptive/ Correlational	Level 4	Utilization of the transition of care tool is able to identify those who will need guarded care and attention for	The evaluation was conducted at two health institutions limiting generalization.

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
Telfair, J., & Smith, W. R. (2016). Self-efficacy and readiness for transition from pediatric to adult care in sickle cell disease. *International Journal of Adolescent Medicine & Health*, *28*(4), 381–388. https://doi.org/10.1515/ijamh-2015-0014	correlates with self-ratings of transition readiness.	sites that were assigned to complete a transition readiness assessment; Transition Intervention Program Readiness for Transition (TIP-RFT).			the transition process. It is recommended that providers use the TIP-RFT tool to measure the overall transition readiness as it will promote effective care coordination.	Findings were limited by measurement as focus was only on self-reported transition readiness.
Whiteman, L. N., Haywood, C., Lanzkron, S., Strouse, J. J., Feldman, L., & Stewart, R. W. (2015). Primary care providers' comfort levels in caring for patients with sickle cell disease. *Southern Medical Journal*, *108*(9), 531–536. https://doi.org/10.14423/SMJ.0000000000000331	To assess the comfort levels of primary care providers caring for patients with sickle cell disease and to identify factors in need of improvement.	Convenient sample of physicians at an annual Johns Hopkins Community Physicians retreat. Survey conducted 19 questions in measuring comfort levels with 4 domains; managing ambulatory care, SCD with other comorbid states, SCD specific issues, and pain management.	Descriptive/ Qualitative	Level 4	A majority of participants lacked confidence with each of the four aspects of caring for individuals with SCD using knowledge gained from residency and medical school. Validation noted in the need for continuing medical education on SCD to ensure that providers are using current information and knowledge.	Study focused on one setting of primary care providers affiliated with one institution limiting generalizability of findings for the general population.
Williams, A. M., & Smith-Whitley, K. (2016). Sickle cell	To promote advocacy to improve care	No sampling. Expert advisory provided on	Descriptive/ Qualitative	Level 7	Proposed benefit in advocacy to improve outcomes of care for	There are valid points presented that can heighten care

Article Title, Author	Study Purpose	Sample	Methods	Level of Evidence	Study Results	Limitations
disease, 2015: A patient advocate's perspective. *American Journal of Preventive Medicine, 51*(1), S5–S9. https://doi.org/10.1016/j.amepre.2016.03.008	practices for individuals with SCD in the United States.	methods to increase access to quality care, institution of partnerships to enhance collaborative care, efforts to improve clinician preparedness and patient self-management.			the patient population of interest however in gains of success support will be needed at institutional, local, state and national levels.	outcomes however there is no definitive means to address funding and sustainability.
Yacoub, M. I., Zaiton, H. I., Abdelghani, F. A., & Elshatarat, R. A. (2019).Effective-ness of an educational program on nurses' knowledge and practice in the management of acute painful crises in sickle cell disease. *Journal of Continuing Education in Nursing, 50*(2), 87–95.	To examine the effectiveness of an educational program on the knowledge and practice of nurses providing care for those in sickle cell crisis.	Purposive sampling of nurses from two hospitals who completed an educational program on sickle cell disease management. Pre-post tests were administered.	Descriptive/ Qualitative	Level 6	Findings indicated a significant difference in nurses' knowledge and care practices after implementation of the educational program.	The study focused solely on nurses not considering the role of other providers likely to be involved in the patients' care. Sample size of nurses was small considering number of RNs working collectively on the hematologic units.

Appendix B

AMSTAR Measurement Tool

Assessment of Multiple Systemic Reviews (AMSTAR)	Assessing Methodological Quality of SRs	11 Domains	1. Was a prior design provided? 2. Was there duplicate study selection and data extraction? 3. Was there a comprehensive literature search? 4. Was the status of publication used as an inclusion criterion? 5. Was a list of studies (inclusion/exclusion) provided? 6. Were characteristics of the studies provided? 7. Was the scientific quality of the included studies assessed and documented? 8. Was the scientific quality of included studies used appropriately to formulate conclusions? 9. Were the methods used to combine the findings of studies appropriate? 10. Was publication bias assessed? 11. Was conflict of interest included?	Scoring of individual Items: 1 point for answers "Yes", "No", "Can't answer", or "Not applicable"

From “Improving the Methodological Quality of Single-Case Experimental Design Meta-Analysis,” by L. Jamshidi, L. Declercq, J. Ferron, M. Moeyaert, S. Beretvas, and W. Noortgate, 2018, *Journal of Mental Health and Clinical Psychology, 2*(4), p. 3.

Appendix C

AMSTAR Quality Appraisal of Included Studies

Adams-Graves & Bronte-Jordan (2016) Quality Score = 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?		*		
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?		*		
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Bernier et al. (2018) Quality Score = 8/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			

Brennan-Cook et al. (2018) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion			*	
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Bulgin et al. (2018) Quality Score= 10/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?	*			
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?		*		
Was the conflict of interest included?	*			

Cramer et al. (2020) Quality Score= 6/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?		*		
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?		*		
Evensen et al. (2016) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?		*		
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			

Freiermuth et al. (2016) Quality Score= 9/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Haywood, Lanzkron, et al. (2015) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?	*			
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?			*	
Was the conflict of interest included?		*		

Haywood, Williams-Reade, et al. (2015) Quality Score= 9/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Jenerette et al. (2016) Quality Score= 8/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?	*			
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?			*	
Was the conflict of interest included?		*		

Kanter et al. (2020) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion		*		
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?		*		
Was the conflict of interest included?	*			
Kayle et al. (2016) Quality Score= 8/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?		*		

Lovett et al. (2017) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?		*		
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?			*	
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?		*		
Was the conflict of interest included?	*			
Matthie & Jenerette (2015) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?				*
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion		*		
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?		*		
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			

Oyedeji & Strouse (2020) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?		*		
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion			*	
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Ross et al. (2021) Quality Score= 10/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?	*			
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			

Singh et al. (2016) Quality Score= 8/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?	*			
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?		*		
Was the conflict of interest included?		*		
Treadwell et al. (2016) Quality Score= 6/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?		*		
Was the conflict of interest included?		*		

Whiteman et al. (2015) Quality Score= 7/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?		*		
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			
Williams & Smith-Whitley (2016) Quality Score= 5/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?		*		
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?		*		
Were the characteristics of the included studies provided?		*		
Was a quality assessment provided and documented?		*		
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?			*	
Was publication of bias assessed?	*			
Was the conflict of interest included?	*			

Yacoub et al. (2019) Quality Score= 9/11				
AMSTAR Question	**Yes**	**No**	**Can't Answer**	**Not Applicable**
Was a prior design provided?	*			
Was there duplicate study selection and data extraction?		*		
Was a comprehensive literature search?	*			
Publication status as an inclusion criterion	*			
Were list of studies (included and excluded) provided?	*			
Were the characteristics of the included studies provided?	*			
Was a quality assessment provided and documented?	*			
Was the scientific quality of the included studies used appropriately in formulating conclusions?	*			
Were methods used to combine findings of studies appropriate?	*			
Was publication of bias assessed?			*	
Was the conflict of interest included?	*			

Appendix D

Institutional Review Board Approval

LIBERTY UNIVERSITY.
INSTITUTIONAL REVIEW BOARD

January 20, 2021

Tracey Royer
Vickie Moore

Re: IRB Application - IRB-FY20-21-525 Improving Clinician Care Practices of Patients with Sickle Cell Anemia

Dear Tracey Royer and Vickie Moore,

The Liberty University Institutional Review Board (IRB) has reviewed your application in accordance with the Office for Human Research Protections (OHRP) and Food and Drug Administration (FDA) regulations and finds your study does not classify as human subjects research. This means you may begin your research with the data safeguarding methods mentioned in your IRB application.

Decision: No Human Subjects Research

Explanation: Your study is not considered human subjects research for the following reason:

(1) "Scholarly and journalistic activities (e.g., oral history, journalism, biography, literary criticism, legal research, and historical scholarship), including the collection and use of information, that focus directly on the specific individuals about whom the information is collected," are not considered research according to 45 CFR 46.102(l)(1).

Please note that this decision only applies to your current research application, and any modifications to your protocol must be reported to the Liberty University IRB for verification of continued non-human subjects research status. You may report these changes by completing a modification submission through your Cayuse IRB account.

Also, although you are welcome to use our recruitment and consent templates, you are not required to do so. If you choose to use our documents, please replace the word *research* with the word *project* throughout both documents.

If you have any questions about this determination or need assistance in determining whether possible modifications to your protocol would change your application's status, please email us at irb@liberty.edu.

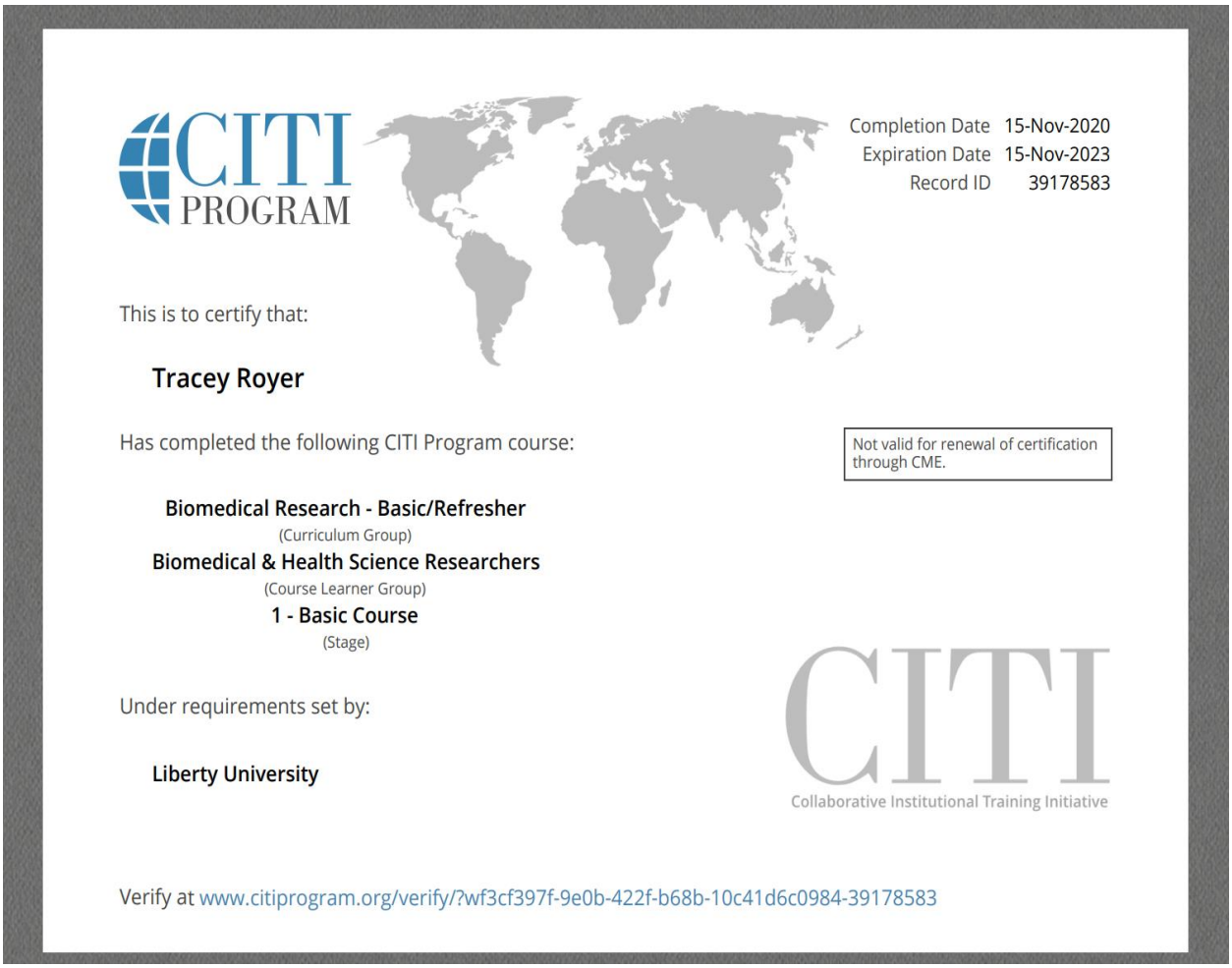
Sincerely,

3/11/2021 Mail - Royer, Tracey - Outlook

G. Michele Baker, MA, CIP
Administrative Chair of Institutional Research
Research Ethics Office

Appendix E

Collaborative Institutional Training Initiative Training Certificate



CITI PROGRAM

Completion Date 15-Nov-2020
Expiration Date 15-Nov-2023
Record ID 39178583

This is to certify that:

Tracey Royer

Has completed the following CITI Program course:

Not valid for renewal of certification through CME.

Biomedical Research - Basic/Refresher
(Curriculum Group)
Biomedical & Health Science Researchers
(Course Learner Group)
1 - Basic Course
(Stage)

Under requirements set by:

Liberty University

CITI
Collaborative Institutional Training Initiative

Verify at www.citiprogram.org/verify/?wf3cf397f-9e0b-422f-b68b-10c41d6c0984-39178583